EUROPEAN STROKE ORGANISATION

FOCUSED WORKSHOP

“Rare Causes of Stroke”

Abstract book
Case reports by participants

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EDITOR: PROF. ANITA ARSOVSKA

NAVIGATING HEMORRHAGIC RISKS IN FABRY DISEASE-A LITTLE-KNOWN TALE

Simonetti Renato[1,2], Pes Federica[2], Laudisi Michele[2], Rodriguez-Luna David[1], De Vito Alessandro[2]

[1]*Vall d'Hebron University Hospital, Stroke Unit, Neurology, Barcelona, Spain*

[2]*Ospedale Universitario "Sant'Anna", UOS Stroke Unit, Ferrara, Italy*

Introduction: Fabry's disease is an X-linked lysosomal storage disorder caused by a deficiency of alpha-galactosidase A (α-Gal A). This deficiency leads to the progressive accumulation of globotriaosylceramide and related glycosphingolipids in the vascular endothelial lysosomes of organs such as the kidneys, heart, skin, and brain[1]. Ischemic stroke is a significant and well-known complication of Fabry's disease and affects over 25% of patients[2]. In those with the classic phenotype, α-Gal A activity levels are extremely low or absent, resulting in the accumulation of glycosphingolipids that cause renal, cardiac, and cerebrovascular issues, ultimately leading to early mortality.[1] Conversely, patients with higher residual levels of α-Gal A activity exhibit less severe symptoms, which may include renal and cardiac variants. An atypical variant of Fabry's disease, presenting primarily with cerebrovascular complications, has also been identified. While cerebral infarction is a well-known consequence of Fabry's disease,[2] intracerebral hemorrhage has not been as widely acknowledged, referring exclusively to case reports or short case series predominantly involving older age groups.[3]

Case presentation: We present the case of a 48-year-old male Fabry patient with a W162R mutation (single base substitution that cause missense mutation; T to C transition in codon 162 of exon 3, predicting a tryptophane-to-arginine replacement[4]), who, over the course of the disease, developed both ischemic and hemorrhagic stroke episodes. Diagnosed with Fabry disease in 1994, the patient has been treated with agalsidase alpha (1 mg/ml) since 2014. Over the course of the disease, the patient developed cardiovascular complications: arterial hypertension treated with a combination of medications (ACE inhibitors, clonidine, beta-blockers, calcium channel blockers, and furosemide), echocardiographic findings of left ventricular hypertrophy, and renal complications: renal failure identified in April 2014 in the uremic phase, necessitating thrice-weekly dialysis. Angio-RM imaging eventually revealed megadolichobasilar anomaly. After three hospitalizations for ischemic stroke and remote symptomatic epileptic seizures between 2014 and 2015, the patient was again admitted to our center due to the sudden onset of speech deficits upon waking. The patient independently sought care at a peripheral emergency department and was subsequently transferred to our facility. An urgent brain CT scan revealed a left capsule-lenticular hemorrhage and another smaller hemorrhagic focus in the left temporal region. Upon admission, the neurological examination showed moderate expressive aphasia, mild paretic dysarthria, and mild right hemiparesis (NIHSS: 7). (Fig.1). Throughout the hospital stay, the standard of care for treating intracerebral hemorrhage was implemented, including strict glycemic monitoring and blood pressure control with a target of <140/90 mmHg in the first 72 hours, despite the drug-resistant hypertension which required multiple therapy adjustments. A follow-up brain CT scan at 96 hours showed substantial stability of the pre-existing hemorrhagic foci, along with the emergence of a small additional focal hyperdensity in the right precentral frontal cortex near the vertex. (Fig.2). Despite the emergence of a new hemorrhagic focus, the patient's clinical condition progressively improved. Upon discharge

after 7 days of hospitalization, mild expressive aphasia remained, characterized by some anomia and phonemic paraphasias, along with mild dysarthria (NIHSS: 2).

Discussion: Cerebral hemorrhagic events in the context of Fabry disease remain poorly delineated, largely due to the disease's rarity and its designation as an outlier among the more prevalent ischemic cerebrovascular conditions. The identification of hemorrhagic lesions in brain regions that seemingly do not share a common pathophysiological mechanism underscores the presence of multiple concurrent triggers as multi-drug resistant hypertension, as seen with capsulo-lenticular hemorrhage and might also be related to the degeneration of small cerebral arteries due to glycosphingolipid accumulation in the vessel walls[5–7], a characteristic complication of Fabry's disease, in cases of hemorrhages in atypical sites. Moreover, the case underlines the increased hemorrhagic risk associated with the administration of antiplatelet therapy, such as acetylsalicylic acid, which the patient was receiving upon admission as part of a secondary prevention strategy from prior hospitalizations. The occasional presence of hemorrhagic events in Fabry patients warrants a more cautious approach to initiating antiplatelet therapy in individuals diagnosed with FD, particularly in the absence of additional atheromatous complications and regardless of the lack of vascular malformations (such as arteriovenous fistulas or aneurysms) observable through imaging studies. Finally, it is imperative to consider the compounded risk presented by chronic terminal renal failure in patients undergoing dialysis; this condition, prevalent in the Fabry disease population, further exacerbates the patient's susceptibility to bleeding[8].

Conclusions: Cerebrovascular complications of FD may be also hemorrhagic and their management present a complex interplay of factors, from the accumulation of glycosphingolipids in cerebral vessels to the challenges posed by antiplatelet therapy and the exacerbated risks associated with chronic terminal renal failure in dialysis patients. This underscores the critical need for a nuanced, patient-centered approach that carefully weighs the unique combination of risks each individual faces. As we navigate these complexities, it becomes increasingly clear that there is no one-size-fits-all solution in the management of Fabry disease. Instead, a deep understanding of the disease's pathophysiology, combined with a vigilant assessment of the potential risks and benefits of therapeutic interventions, is essential.

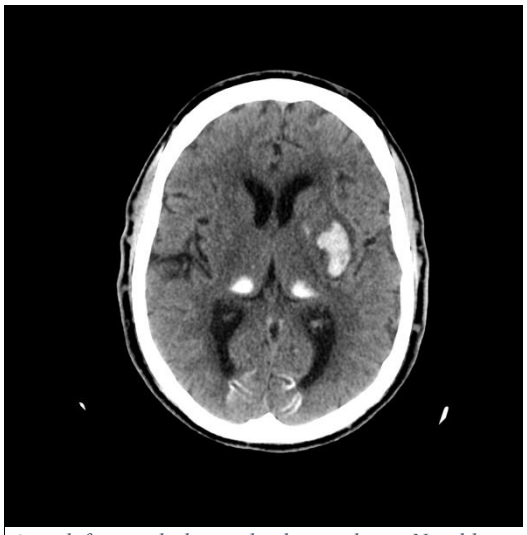

Fig.1: left capsule-lenticular hemorrhage. Notable bilateral "pulvinar sign"

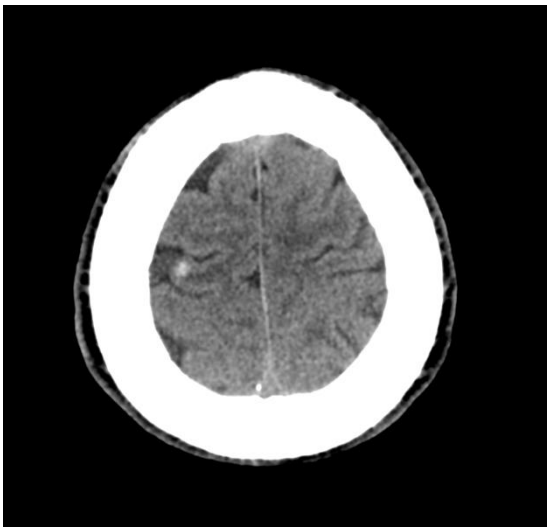

Fig.2: small hemorragic focus in the right precentral frontal cortex near the vertex

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MOYA-MOYA DISEASE AS A RARE CAUSE OF ISCHAEMIC STROKE IN YOUNG FEMALE PATIENT – CASE REPORT

Milovan Roganovic[1,3], Biljana Radovanovic[2], Zeljko Zivanovic[1,2]

[1] Faculty of Medicine, University in Novi Sad, Novi Sad, Republic Serbia
[2] Clinic for Neurology, University Clinical Centre of Vojvodina, Novi Sad, Republic Serbia
[3] Clinic for Neurology, Clinical Centre of Montenegro, Podgorica, Montenegro

Introduction: Moya-moya disease (MMD) is a rare progressive cerebrovascular disease that is characterized by stenosis of the vascular branches of the internal carotid artery and the development of abnormal, fragile collateral vessels. Patients with MMD mainly present with signs and symptoms of cerebral ischaemia or cerebral hemorrhage with a typically progressive course. Although the pathophysiology of this disease is still not clear enough, recent knowledge enables faster diagnosis of it, as well as a modern treatment approach (drug and/or surgical).
Case presentation: Our patient, at that time a 30-year-old pregnant woman, noticed the first symptoms in the form of occasional numbness of the right arm and leg in the second and third trimesters of her second pregnancy. Arterial hypertension was verified by the primary health-care physician and gynecologist, and antihypertensive therapy was introduced. Two days after delivery (June 2019), she developed a mild right-sided sensory symptoms and mild dysphasia (NIHSS 3) and was hospitalized in our center for additional diagnostics and treatment. MRI detected acute ischaemic lesions in the left middle cerebral artery (ACM) vascular territory (nc. caudatus, upper and middle frontal gyrus, postcentral gyrus, cortex/subcortex of the upper temporal gyrus…) as well as several foci in the white matter which correspond to acute ischaemia in the region of the inner "watershead zone". MRA indicate (sub) occlusion of the

M1 segment of left ACM, with collateral distal filling of the M2. The patient underwent an extensive cardiological and laboratory examination according to the protocol for young patients with a thrombotic event - no significant deviations were detected. Acetylsalicylic acid was prescribed. The patient was not motivated for further diagnostics, so she was monitored on an outpatient basis - yearly MRIs were performed, which did not find significant deviations until 2022, when episodes of bilateral tinnitus occurred. The control MRI/A performed at that time showed the progression of the stenotic lesions. This motivated patient to accept further diagnostics, and at the beginning of 2023, a digital subtraction angiography (DSA) was performed, which indicated stenotic lesions of both ACMs, predominantly on the left. At the beginning of 2024, the patient underwent surgical intervention (direct revascularization using extra-intracranial bypass from the superficial temporal artery to a cortical branch of the ACM) and surgical treatment on the right side was also recommended.
Discussion and Conclusion: We presented a young patient with a rare cause of stroke, atypical for the patient's country of origin. Although, according to the patient's choice, the implementation of the gold diagnostic standard (DSA) and thus the definitive diagnosis was delayed, modern treatment approach was applied (best medical treatment and surgery) which should prevent the occurrence of recurrent stroke events.

RECURRENT CEREBROVASCULAR EVENTS IN AN UNTREATED HIV-POSITIVE PATIENT A COMPLEX CLINICAL DILEMMA

Giorgi Matchavariani

Acad. N.Kipshidze Central University Hospital, Tbilisi, Georgia

Introduction: This case presents a 46-year-old HIV-positive male with a striking history of sequential strokes, first ischemic in December 2022 and later hemorrhagic in March 2023. Despite enrollment in the HIV program since 2018, the patient had not received treatment until the recent discovery, prompting exploration into the potential connection between untreated HIV and recurrent cerebrovascular events.
Case Presentation: In March 2023, the patient exhibited symptoms suggesting a prior stroke, including headaches, speech difficulties, and right-sided limb weakness. Imaging revealed softening foci in vital brain regions. After initial stabilization and discharge, a recurrence of symptoms in April 2023 led to the diagnosis of ventricular hemorrhage, lacunar leukomalacia, and cortical atrophy. Despite medical efforts, the patient's condition rapidly deteriorated, culminating in respiratory failure and cardiac arrest on May 12, 2023. EEG and imaging illustrated abnormal brain activity and persistent hemorrhage.
Conclusion: This case underscores the urgency of early antiretroviral therapy in HIV-positive individuals, emphasizing the intricate link between untreated HIV, endothelial dysfunction, and susceptibility to both ischemic and hemorrhagic strokes. The delayed initiation of treatment accentuates the critical importance of timely intervention. Further research is essential to unravel the complex pathophysiological mechanisms governing HIV-associated cerebrovascular complications, providing insights for optimized clinical management and prevention strategies.

LEFT ATRIAL MYXOMA AS A RARE CAUSE OF ISCHEMIC STROKE – CASE REPORT

Stefan Bojovic[1,3], Tamara Rabi Zikic[1,2], Zeljko Zivanovic[1,2]

[1] Faculty of Medicine, University in Novi Sad, Novi Sad, Republic Serbia
[2] Clinic for Neurology, University Clinical Centre of Vojvodina, Novi Sad, Republic Serbia
[3] Clinic for Neurology, Clinical Centre of Montenegro, Podgorica, Montenegro

Introduction: Acute ischemic stroke is frequently attributed to cardioembolism, accounting for approximately 20% of cases. However, within this spectrum of cardiac etiologies, cardiac myxomas (CM) represent a distinct and comparatively rare entity, constituting only 0.5% of cases. Originating predominantly from the left atrium (75%), these tumors exhibit a high predilection for embolic events (35%), particularly targeting the central nervous system and retinal arteries. Compared to tumor tissue itself, adherent thrombus is a more common cause of embolization. Following diagnosis, urgent measures such as vascular recanalization and CM resection are required.

Case Report: A 65-year-old, right-handed female, with a complex medical history including hypothyroidism, Sjogren syndrome, inflammatory bowel disease, pheochromocytoma operation, colorectal cancer following polypectomy ten years prior, and varicose vein surgery, presented to the emergency department with a sudden onset of slurred speech and left-sided weakness, which began approximately 30 minutes before to arrival. She denied any loss of consciousness or headache prior to the symptoms. On initial assessment, she was hemodynamically stable with normal vital signs. Upon neurology examination, the patient exhibited left central facial nerve palsy, dysarthria, and left-sided hemiplegia with left-sided hypoesthesia. The National Institutes of Health Stroke Scale (NIHSS) score was 12. Non-contrast brain CT scan revealed no abnormalities. However, CT angiography of the head and neck revealed thrombosis in the distal M1 segment of the middle cerebral artery (MCA), and proximal M2 segment of the MCA, measuring 13mm in length, accompanied by a well-developed collateral network of blood vessels. Intravenous thrombolysis (IVT) was performed using alteplase at the maximal dosage of 90 mg. Following the infusion, the patient's NIHSS score improved to 0, obviating the need for mechanical thrombectomy. MRI of the brain revealed acute ischemic stroke involving the genu and posterior limb of the right internal capsule, as well as the right thalamus. During hospitalization, additional investigations were conducted to ascertain the etiology of the ischemic stroke. Holter ECG monitoring, carotid artery duplex scan, and TCD Bubble test were normal. Transthoracic echocardiography revealed an echo formation in the left atrium, while transesophageal echocardiography identified an oval, homogeneous echo formation in the left atrium measuring 1.3 x 1.6 cm, attached to the interatrial septum with a wide base, suggestive of atrial myxoma. The patient underwent surgical resection of the cardiac tumor, confirming the diagnosis of atrial myxoma upon histopathological examination. During an eleven-month follow-up period, the patient remained free from symptomatic intracranial hemorrhage (sICH) as well as new ischemic events.

Discussion and Conclusion: We presented a female patient with acute ischemic stroke secondary to the left atrial myxoma that achieved good clinical outcome after IVT. Since IVT was successful in treating the patient, it can be assumed that the thrombus was the cause of the embolus rather than the tumor tissue particles themselves. In our case, IVT was performed before the echocardiographic findings, so CM was not known at the time of thrombolysis. Cerebral angiography is recommended before IVT to rule out myxomatous

aneurysms and reduce the risk of intracranial hemorrhage. Further studies with a larger sample are necessary to provide more evidence on the safety and efficacy of IVT as well as bridging therapy for the treatment of CM-ischemic stroke patients. Currently, there is low-grade evidence for therapeutic strategies, mostly based on case reports or small case series.

CEREBRAL AMYLOID ANGIOPATHY - RELATED INFLAMMATION: A CASE REPORT

Ivana Karla Franić

Department of Neurology, University Hospital Centre Zagreb, Zagreb, Croatia

Introduction: Cerebral amyloid angiopathy-related inflammation (CAA-ri) is a rare entity that lies on a spectrum between CAA and vasculitis, characterized by autoimmune neuroinflammatory response against beta-amyloid protein deposits in leptomeningeal and small cortical vessels.[1,3]
Case presentation: We report a 65-year-old male patient who was hospitalized twice due to repeated seizures. Both times MSCT showed recurrent right frontoparietal subarachnoid hemorrhage (SAH). Initial MRI showed suspected meningoencephalitis. MRI one month later described an extensive vasogenic edema (VE) in right temporoparietal white matter; sulcal SAH; hemosiderin deposits; acute ischemia in right parietal cortex and cortical superficial siderosis. Clinically he reported only cognitive impairment. Intravenous pulse corticosteroid therapy (PCST) was administered. Control MRI showed complete regression of VE. One month later he had a relapse in form of occasional transient stereotype left cheek paresthesia and cognitive decline. MRI showed reactivation of the inflammatory process in the left frontoparietal region and PCST was repeated. VE completely regressed on control MRI. During the follow up period, MRIs show stationary intracranial findings. Patient is clinically stable without new neurological deficits.
Discussion: CAA-ri shares neuroradiological characteristics with CAA,[1,2,3] but differs in asymmetric cortico-subcortical T2 white matter hyperintensities (WMH),[1,3] which can produce a mass effect in form of apparent diffusion, suggestive of vasogenic edema.[1,3] Definite diagnosis is made via biopsy.[1,3] CAA-ri responds well to high doses of corticosteroids.[1,2,3]
Conclusion: Despite the relapsing clinical course, an excellent therapeutic response to each cycle of corticosteroids shows the importance of timely recognition, and comprehensive and persistent approach in the treatment of CAA-ri.

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FIBRINOGEN DEGRADATION COAGULOPATHY FOLLOWING INTRAVENOUS THROMBOLYSIS IN A PATIENT WITH STROKE

Aurelija Daškevičiūtė [1], Vitalija Šumskienė[1]

[1]Center of Neurology, Vilnius University, Vilnius, Lithuania

Introduction: Fibrinogen depletion is a potential cause of intracranial hemorrhage following intravenous thrombolysis (IVT). We present a case of fibrinogen degradation coagulopathy following IVT.
Case presentation: A 67-year-old man presented with rightward eye deviation, left-sided facial prosoparesis and left arm weakness with a NIHSS score of 5, within 2.5 hours from symptom onset. Head CT-angiography with perfusion imaging revealed an 11.5 mm thrombus within the M2 segment of the right middle cerebral artery, resulting in a perfusion deficit measuring 246 cm^3 and an ischemic core of 49 cm^3 with an ASPECTS score of 9. The patient had no contraindications and received Alteplase in accordance with his weight and was transferred for mechanical thrombectomy. Full reperfusion was achieved during a single-session mechanical thrombectomy. 4 hours after IVT the patient deteriorated significantly with a NIHSS score of 14. Head CT revealed ~143 cm^3 right fronto- parietal intracerebral hemorrhage penetrating into the ventricles and subarachnoid space. Additionally, epistaxis and hematuria started. The red blood cell count was 3.17 x 10^{12} g/l, platelet count 95 × 10^9/L, fibrinogen level <0.4 g/l, and d-dimers 17540 (µg/L). In response to coagulopathy, the patient received 20 units of cryoprecipitate, 2 g of fibrinogen concentrate, and 2 units of platelets. Following the reversal of coagulopathy, craniotomy and hematoma removal were performed. The patient demonstrated no neurological improvement, ultimately leading to death.
Discussion and conclusion: Routinely measuring fibrinogen levels before and after intravenous thrombolysis may be beneficial for identifying patients at high risk of fibrinogen depletion coagulopathy and post-thrombolytic intracerebral hemorrhage.

A RARE CASE OF GENETIC UNBALANCED TRANSLOCATION PRESENTING AS INCOMPLETE HANAC SYNDROME, SEIZURES AND LEARNING DIFFICULTIES

Simina Cosma

Manchester Centre for Clinical Neurosciences - Stroke department, Salford Royal Hospital Salford, United Kingdom

Background and aims: COL4A1/A2 genes encode type IV collagen, a key component of the basement membrane of vascular endothelium. Mutations of COL4A1/A2 genes result in multi-system disorders. The principal phenotype described in adults is HANAC syndrome (hereditary angiopathy with nephropathy, aneurysms and muscle cramps). More than 100 mutations have been described, half of which are inherited in an autosomal dominant pattern, the rest being de novo mutations.
Methods: A 40-year-old lady with a history of learning disabilities and seizures presented with accelerated decline in cognition and function in her 3rd decade of life. MRI brain and MR angiogram revealed extensive white matter changes, microhemorrhages and old lacunar infarcts consistent with cerebral microangiopathy, alongside three aneurysms involving the middle cerebral artery and its branches. Renal function and creatine kinase were normal.
Results: DNA microarray analysis identified a gain of 59 coding proteins in chromosome 13

involving the COL4A1 and COL4A2 genes (chromosome 13 duplication). A terminal deletion in the short arm of chromosome 8 was identified with an interstitial gain adjacent to the deletion (8p inverted duplication and deletion syndrome). Parental karyotyping proved this to be a de novo mutation.
Conclusion: We report a case of an unbalanced chromosomal translocation occurring de novo affecting the COL4A1/A2 genes resulting in incomplete HANAC syndrome. The co-existent chromosome 8 mutation led to the additional features of seizures and learning disabilities. To our knowledge, this particular genetic translocation between chromosomes 13 and 8 with this resultant clinical phenotype has not been reported before.

MOYA MOYA DISEASE

Miguel Trindade

Stroke Unit, Hospital Professor Doutor Fernando Fonseca, Lisbon, Portugal

Case presentation: A twenty-eight-year-old male patient was admitted to our stroke unit with numbness of the left side of his face and of the left upper limb, in which he also noticed weakness in the hand. He recalled having this numbness since 2019 and always while doing cardio or heavy-weight training in the gymnasium. More specifically, he recalled having this feeling only on the third of three repetitions when making maximum effort. In the previous episodes, he never noticed weakness until the day of admission. He has been a smoker (stopped five years before) but still smoked cannabinoids. The patient hadn't had a drink in the last two years. He wasn't taking any medication, including steroids, herbal or recreative drugs. On admission, he had a blood pressure of 133/69mmHg and glycemia of 98mg/dL, and the neurologic exam was remarkable for a minor left facial palsy and a distal weakness in the left hand with impairment of fine motor skill (like writing) - NIHSS 1. He underwent CT with angiography that showed a small acute hematoma in the right lenticulo-capsular region and bilateral substantial narrowing of the middle cerebral arteries with hyperplasia of the lenticulo-striate arteries. That angiographic pattern was highly suggestive of Moyamoya syndrome, and he was admitted to our stroke unit. In our stroke unit he underwent subsequent diagnostic workup for moyamoya syndrome (shown in ppt presentation):Carotid and vertebral ultrasonography showed reduced pulsatility of the right internal carotid artery, which was compensated by increased pulsatility in the ipsilateral external carotid artery; increased velocities and flows in the vertebral circulation, particularly in the right vertebral artery; intima-media thickness (IMT) was normal and atherosclerotic plaques were absent. Diagnostic angiography showed steno-occlusive disease in the right internal carotid artery with no flow in the M1 segment of the middle cerebral artery (MCA) and pre-occlusive stenosis of the A1 segment of the anterior cerebral artery (ACA); adequate flow in the left internal carotid artery with steno-occlusive disease of the ACA (A1 and A2) and complete occlusion of the left MCA (M1); bilateral hypertrophy of the lenticulo-striate arteries with a puff of smoke configuration, compatible with Moyamoya angiographic pattern; there is compensation from the right vertebral artery to anterior circulation in both hemispheres, particularly the right one; steno-occlusive disease of the P1 segment of the left posterior cerebral artery (PCA). These findings are classified as Suzuki grade III on the right and grade II on the left. Magnetic resonance imaging (MRI) showed the small right lenticulo-striate hemorrhagic lesion and bilateral cortical frontal, parietal, temporal, and insular ischemic acute

and subacute lesions; occlusive stenosis of the right MCA, steno-occlusive stenosis of the left MCA, and bilateral hypertrophy of the lenticulo-striate arteries. Perfusion MRI showed diminished regional cerebral blood flow (rCBF) and increased mean transit time in the right MCA territory. Hemoglobin electrophoresis was normal, no dyslipidemia nor diabetes was found, and the autoimmune workup was also negative.No kidney disease (such as polycystic disease) was found on ultrasonography, and the doppler of the renal arteries was also unremarkable. The patient was proposed and accepted for surgical revascularization and was discharged with almost complete recovery of the weakness in the left arm.

BRIGHTENED VESSELS SHEDDING LIGHT ON THE GREAT IMITATOR. A CASE REPORT OF VESSEL WALL MRI IN SYPHILITIC CEREBRAL VASCULITIS

Del Bello Beatrice, MD

Department of Brain and Behavioral Sciences, University of Pavia, Italy
Department of Stroke Unit and Emergency Neurology, IRCCS Mondino Foundation, Pavia

Case presentation: A 48-year-old man was admitted to the Department of Emergency Neurology and Stroke Unit of our hospital after an acute episode of severe gait instability occurred ten days earlier. He was first admitted to the Emergency Department (ED) of another hospital for a fall consequent to the gait disturbance, resulting in a left distal radius fracture. On that occasion, blood pressure was 220/110 mmHg and he was treated with a bolus of Urapidil. Neuroimaging was not performed, and he was discharged home after blood pressure control. However, in the next week the symptoms persisted, and he was admitted to our hospital for further evaluation. The patient's neurological symptoms began six months earlier with mild gait disturbance and cognitive impairments. The symptoms progressively worsened in the following months, and after about two months he was unable to walk unassisted. He first started to use a cane, but soon after he started to use crutches. For this reason, he went to live with his brother a month before admission. Past medical history was relevant for arterial hypertension, a previous episode of major depression, 20 pack-year smoking history, and alcohol abuse. He drank an average of two liters of wine per day, but he had stopped few weeks earlier. He also reported occasional cocaine consumption. He denied family history of neurologic disorders. On hospital admission, the patient was alert and partially oriented in time and space. Fluency was scarce with moderately dysarthric speech, while naming and comprehension were unremarkable. Cranial nerves examination revealed horizontal saccadic dysmetria. There were no objectifiable motor deficits. Deep tendon reflexes were brisk with left side prevalence. He had an upgoing left plantar response. Sensation was difficult to examine because of the patient's lack of collaboration, but no evident sensory deficits could be detected. The finger-to-nose and heel-to-shin tests revealed mild bilateral dysmetria. Gait was wide-based. General examination was normal. He denied headaches or systemic symptoms. Screening of neuropsychological evaluation with Mini Mental State Examination revealed severe cognitive impairment (score: 8/30). Patient's blood tests (complete blood count, electrolytes, liver and kidney functions, blood glucose) were within normal limits. C-reactive protein was 3,1 mg/dL (normal < 0,5), total cholesterol was 163 mg/dL, and LDL-cholesterol was 129 mg/dL. ECG showed sinus rhythm and chest X-ray was normal. NCCT showed severe leukoaraiosis involving both subcortical and deep white matter bilaterally, and mild bilateral frontal and cerebellar atrophy. Magnetic Resonance Imaging (MRI) of the brain

confirmed the extensive white matter lesions seen on NCCT and revealed multiple acute subcortical ischemic lesions in supratentorial regions (Figure 1). Time-of-flight (TOF) MR angiography showed a diffuse irregular profile of the intracranial vessels without moderate-severe intracranial artery stenosis (Figure 2). Considering the multiterritorial ischemic lesions and the clinical picture, to better define stroke etiology a vessel wall MRI (vwMRI) study was performed. It showed the presence of multifocal concentric contrast enhancement of the major intracranial arteries, involving the carotid syphons, the proximal segments of the middle cerebral artery (MCA) bilaterally and the left posterior cerebral artery (PCA), the proximal third of the basilar artery, and the intracranial segments of the vertebral arteries (Figure 3). These findings were suggestive for a vasculitis involving the cerebral arteries[1], so a lumbar puncture was performed.Cerebrospinal fluid (CSF) analysis showed an albumin concentration of 53 mg/dL (normal 10-30), mild blood-brain barrier disruption (albumin CSF:serum ratio of 1,5%, normal < 0,70), normal glucose, and no pleocytosis. Beta-amyloid, tau and phospho-tau levels were normal. To define the etiology of the ongoing inflammatory process, a full infectious and autoimmune screening was performed. Serum autoimmune panel including antinuclear antibodies, extractable nuclear antigen panel, anti-neutrophil cytoplasmic antibodies, rheumatoid factor, anti-cyclic citrullinated peptide, anti-double stranded DNA, complement C3 and C4, Lupus anticoagulant, anticardiolipin, and anti-beta2 glycoprotein I antibodies was unremarkable. Infectious workup was negative for HBV, HCV, and HIV. Serum Treponema Pallidum Hemagglutination test (TPHA) and Rapid Plasma Reagin (RPR) tests were both positive. Treponemal-specific antibody testing on CSF was then performed but yielded negative results. Even though the absence of CSF-Treponemal antibodies is used to rule out neurosyphilis[2], in the setting of a very high clinical suspicion with a supportive serum antibody testing (with positivity of both treponemal and non-treponemal tests) and compatible neuroimaging findings, as occurred in our case, the diagnosis of a syphilitic vasculitis should not be excluded.[3] Considering the neurological picture, serology and vwMRI findings, a diagnosis of syphilitic CNS vasculitis[4] and treatment was started accordingly with intravenous penicillin and prednisone (1 mg/kg/die) was started. To better define the systemic involvement of the pathology, the patient underwent CT-angiography of the supra-aortic trunks, intracranial arteries, chest, abdomen, and lower extremities. CTA confirmed the presence of mild multifocal stenosis of the intracranial arteries alternated with mildly dilated segments; these findings were compatible with the diagnosis of cerebral vasculitis. The rest of the exam revealed widespread atherosclerotic alterations in both splanchnic and limbs arteries, without findings suggestive for a vasculitis involvement of other vessels. Atorvastatin 40 mg and acetylsalicylic acid 100 mg were added based on the CTA findings. Neurological examination after two weeks of intravenous antibiotic treatment showed mild improvement of the ataxic gait despite persisting severe cognitive deficits. Prednisone was tapered over the next weeks. The patient was then transferred to a long-term care facility. Only telephonic follow-up is available. The patient's brother reported no new acute neurological symptoms in the following three years. Despite a mild improvement on motor deficits and gait instability, his cognitive impairment remained severe. The patient still need assistance, and he was not able to return living at home.

Figure 1.

Diffusion-weighted imaging (DWI) shows several subcortical lesions with restricted diffusion at the level of the right parieto-occipital (A, arrow) and left thalamic (A, arrowhead), right posterior temporal lobe (B), and bilateral frontal lobes (C). T2 fluid-attenuated inversion recovery (FLAIR) sequence demonstrates diffuse white matter hyperintensities (D, E, F).

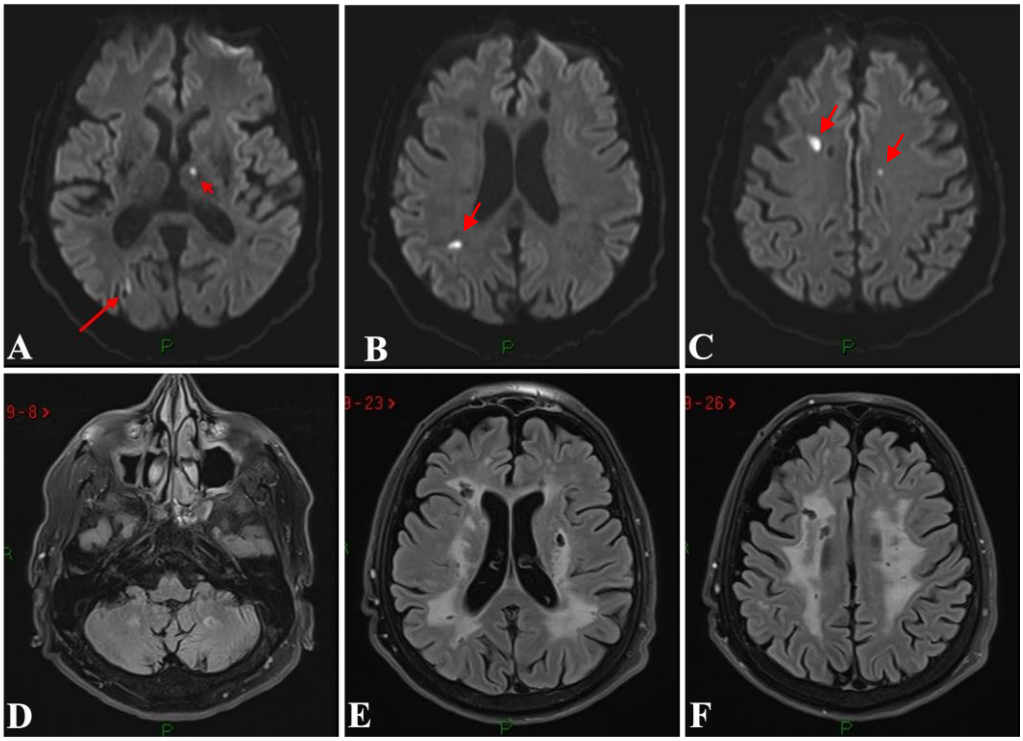


Figure 2.
Time-of-flight (TOF) MR angiography shows multiple parietal irregularities of intracranial vessels.

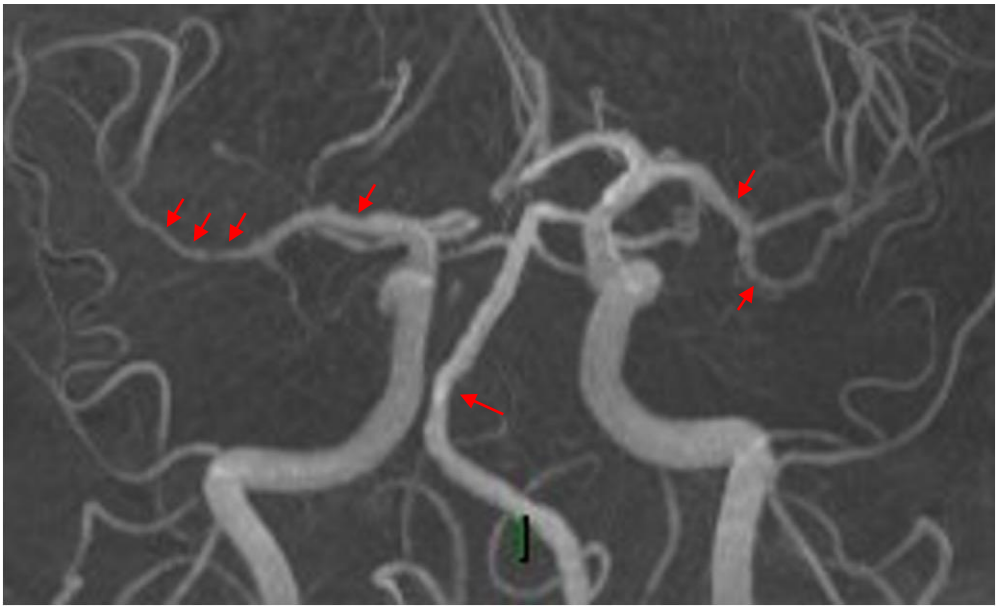
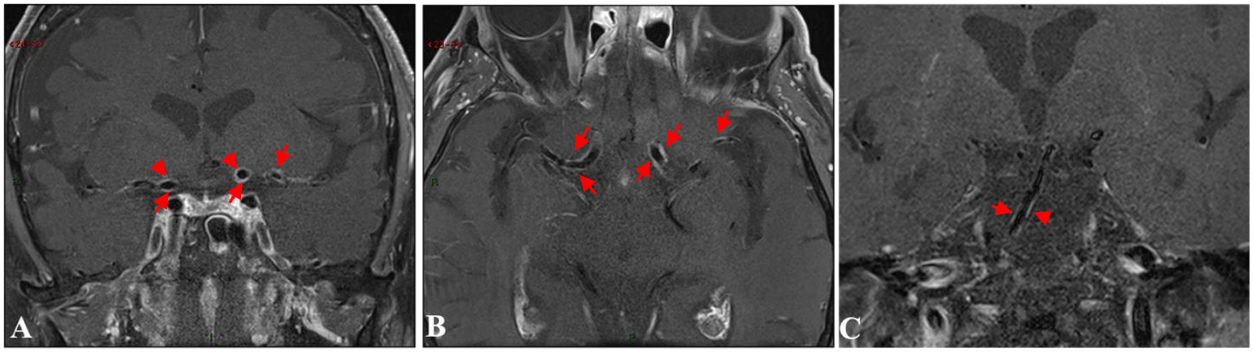


Figure 3.

Black-blood T1-weighted sequences after Gadolinium administration show concentric contrast enhancement of the intracranial arterial wall, involving both anterior (A, B) and posterior circulation (C).



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CNS VASCULITIS AND STROKE

Vojtech Novotny

Department of Neurology, Haukeland University Hospital, Bergen, Norway

Introduction: Paraneoplastic vasculitis of the central nervous system (CNS) is a rare condition that can occur alongside specific types of cancer. The exact pathophysiology is not fully understood, but an immune response leading to a prothrombotic state and affecting the vascular walls is probably involved. An ischemic stroke is often the initial clinical manifestation of CNS vasculitis. Ischemic lesions are often small, disseminated in time, and involve multiple arterial territories. Therefore, the clinical picture may present as acute or eventually subacute encephalopathy or multiple clinically distinct transient ischemic attack (TIA) episodes.
Case presentation: I present a male in his 60s who was admitted several times to the stroke unit with TIAs and small brain infarctions in multiple arterial territories. Computer tomography angiography, on later admissions, revealed several segmental stenoses in various small and medium-sized cerebral arteries. The stenoses were initially treated with Nimodipine under close monitoring with transcranial ultrasound, as multiple vasospasms were suspected. However, the stenoses persisted despite the treatment. The clinical picture therefore

suggested CNS vasculitis as a likely cause of recurrent cerebrovascular events. A very good response to initiated immunosuppressive therapy with prednisolone supported this diagnosis. Further investigation ruled out systemic vasculitis but revealed cervical lymphadenopathy. It was initially investigated with a fine needle biopsy that yielded negative results, but subsequent surgical biopsy confirmed the presence of classical Hodgkin's lymphoma. It appeared that the paraneoplastic CNS vasculitis secondary to Hodgkin's lymphoma was responsible for the patient's recurrent cerebrovascular events. Treatment targeting the cancer itself, along with secondary prevention using immunosuppression and low molecular weight heparin, proved to be effective with no recurrent stroke thereafter. Transcranial ultrasound was an important part of the diagnostic work-up but also monitoring of treatment response. After Hodgkin's lymphoma was cured, no recurrent TIAs or ischemic strokes were reported.

LOCKED-IN SYNDROME DUE TO SEVERE CASE OF VARICELLA ZOSTER VIRUS VASCULITIS

Triin Helin Unt

West-Tallinn Central Hospital, Neurology and Psychiatry Clinic, Stroke department, Tallinn, Estonia

Introduction: Varicella zoster virus (VZV) vasculitis is a rare but serious complication arising from the reactivation of the varicella zoster virus. This condition primarily affects the elderly and immunocompromised individuals, posing significant challenges in diagnosis and management. The virus can lead to vasculitis by directly invading the blood vessel walls or triggering an immune response that targets vascular structures. VZV vasculitis can have diverse clinical presentation, ranging from skin lesions and neurological deficits to systemic manifestations. Skin findings often include vesicular eruptions in a dermatomal distribution, resembling herpes zoster, while neurological involvement may lead to cerebrovascular accidents or encephalitis. The intricate interplay between viral factors and the host's immune response contributes to the complex pathogenesis of VZV vasculitis.
Case presentation: This case presentation demonstrates a 69-year-old female patient with severe course of VZV-vasculitis which first presented with headache, meningismus, undulating dysartria and general malaise but over next week progressed to locked-in syndrome due to vast ischemic damages in brainstem and cerebellar region, shown on images 1-3. Diagnosis was confirmed by radiological findings and presence of VZV IgG antibodies in cerebrospinal fluid. The patient was treated with intravenous acyclovir for 21 days, along with pulsed treatment with intravenous methylprednisolone, without remarkable positive clinical effect.

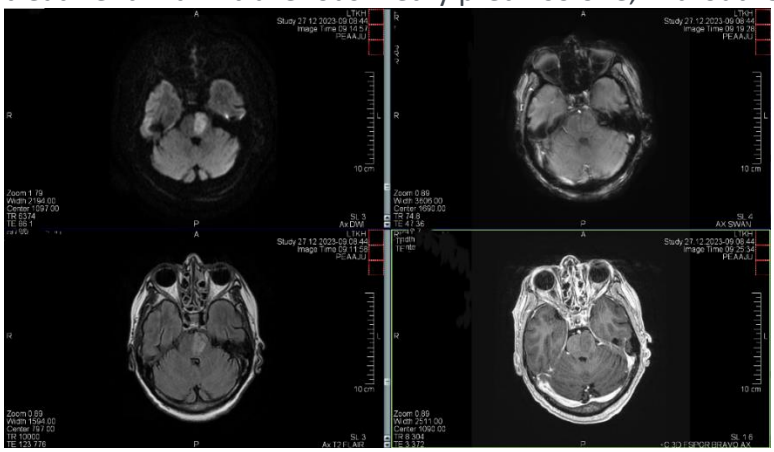

Image 1 Second MRI scan showing ischemic lesion in the brainstem on the left.

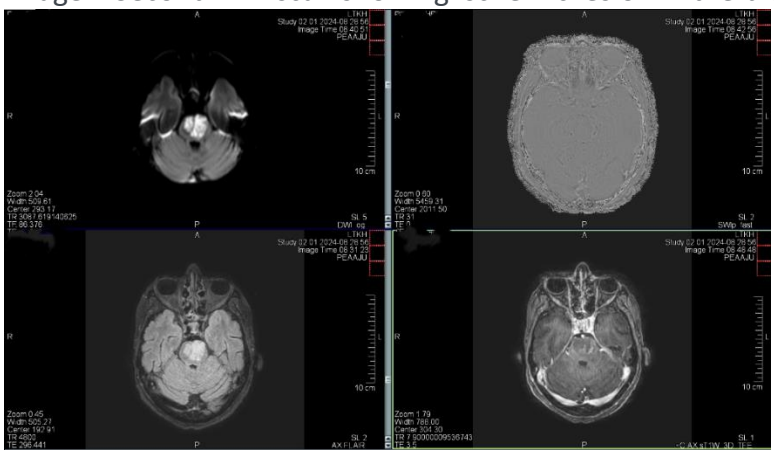

Image 2 Third MRI scan showing ischemic lesion's prgression in the brainstem.

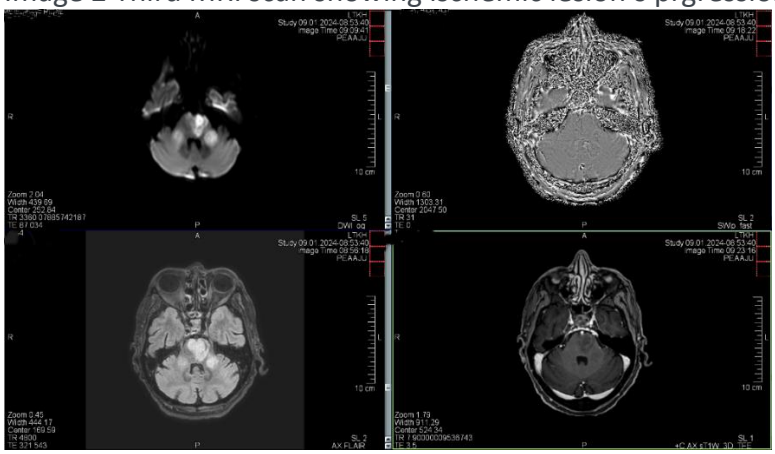

Image 3 Fifth MRI scan showing ischemic lesions' dynamic in the brainstem and cerebellar peduncles.

Conclusions: Diagnosing VZV vasculitis requires a multidisciplinary approach, combining clinical evaluation, imaging studies, and laboratory analyses. Early recognition is crucial to initiate prompt antiviral and immunosuppressive therapy, aiming to mitigate the inflammatory response and prevent further vascular damage. This abstract emphasizes the need for awareness among clinicians, given the potential for severe morbidity and mortality associated with VZV vasculitis. Improved understanding of pathogenesis and advancements in diagnostics will enhance patient outcomes in the future.

Acknowledgment
Thank you to my collegues at West-Tallinn Central Hospital - Karin Kannel, Sandra Ütt, Ulvi Thomson, Ain Vares, Alo-Rainer Leheste, Sandra Marii Mallene, Katrin Gross-Paju

ERYTHREMIA AND STROKE

Nika Kvernadze

Neurology/Neurosurgery Department, Stroke Center
K. Eristavi National center of experimental and clinical surgery, Tbilisi, Georgia

Case presentation: 68-year-old man. Clinical presentation: disoriented; difficulty speaking, NIHSS-4. The face was significantly hyperemic. He was hospitalized in the clinic about 6 hours later time onset of symptoms. MRA scan - without damage. According history: Arterial hypertension, erythremia, (diagnosed 21 years ago), ischemic stroke 5 yesrs ago (without residual defect), thrombosis of the deep veins of the lower limbs, no AF, no carotid artery stenosis. Treated - Rivaroxaban 20 mg; Aspirin 100mg; Hydroxycarbamide 500 mg and with Antihypertensive drugs. According to the laboratory analysis, attention was drawn to a pronounced plethora, Hemogram: RBC-6.37 10^12/l, HGB-21.2g/dL, HCT-65.7%, PLT-481 10^9/l. The patient was placed in the intensive care unit of the stroke center. Based on the anamnesis and laboratory analyses, patient was consulted by a hematologist. On his recommendation, erythrocytopheresis was performed. After erythrocytopheresis procedure – was checked CBC. According to the general analysis of control blood, RBC-5.58 10^12/l, HGB-19.2 g/dL, HCT-58.2 %, PLT-395 10^9/l. After his procedure. the patient's condition improved, the neurological deficit regressed and returned to the initial position, The degree of hyperemia on the face also decreased. On the recommendation of the hematologist, Ruxolitinib 20 mg was added to the treatment.

STROKE AS AN EARLY COMPLICATION IN BACTERIAL MENINGITIS

Ciopleias Bogdan -Nicolae

Faculty of Medicine, Transilvania University Brasov; Department of Neurology, County Clinic Hospital, Brasov,Romania

Introduction: Systemic infections have been associated with an increased risk of stroke, inflammation being considered the main risk factor. Delayed cerebral thrombosis (DCT) is a cerebrovascular phenomenon that can be encountered in patients that recover form pneumococcal meningitis. The aetiology is not clear, direct bacterial invasion, activation of coagulation, endothelial dysfunction and inflammation being suggested.
Case Presentation: A 47-year-old male, without any medical history or risk factors, presented in the emergency department for acute confusional state and sever psychomotor agitation, superposed on high fever. 3 days before admission the patient presented rhino-pharyngitis symptoms for which received medical consultation from the family doctor. At the neurological exam, severe disorientation, positive Budzinski sign, positive Kernig sign, without other focal neurological deficits. Native CT scan showed no pathological findings, contrast enhanced cranio-cerebral CT showed a superior sagittal sinus thrombosis. Blood count showed severe inflammatory syndrome and lumbar puncture revealing very high protein and leucocytes levels and low glucose, microbial culture being positive for Streptococcus pneumoniae. Antibiotherapy and LMWH in therapeutic dosage was started with rapid improvement of the symptoms. During the 9^{th} day of hospitalization, the patient became somnolent developing left oculomotor and trochlear nerve palsy, bilateral abducens neve palsy and mild right hemiparesis. The MRI scan showed acute ischaemic lesions in the left pontine and cerebellar peduncle regions, right occipital periventricular region and left temporal subcortical region.
Conclusions: DCT is a rare complication influencing the outcome of bacterial meningitis patients. This case presentation reports the development of stroke following bacterial meningitis, a possible pathophysiological mechanism being the delayed inflammatory response which lead to diffuse endothelial injury and thrombosis.

POST – RADIATION CEREBRAL NECROSIS

Marta Jeremić

Neurology Clinic, Clinical Center of Serbia, Belgrade, Serbia

Case presentation: We present a 44 year old male patient. He is unemployed and unmarried. By profession he is a mechanical engineer. His initial neurologic symptoms were drowsiness, confusion and disorientation. The symptoms began about 10 days before he was admitted to hospital. He was complaining of slurred speech, confusion and disorientation. He showed signs of altered mental state and changes in personality. He also had urinary urgency followed by occasional urinary incontinence. Because of these symptoms he was admitted to the Neurology Clinic, Clinical Center of Serbia. He had the following vascular risk factors: hypertension, hyperlipidemia, hyperhomocysteinemia, and he was a smoker. In his medical history we have obtained information that in 2011 he had an ischemic stroke in the vascular territory of the right medial cerebral artery. An ischemic lesion was present on neuroimaging (pre-and postcentrally frontoparietally on the right). He had a residual neurologic deficit manifesting as a slight left sided spastic hemiparesis and a moderate dysarthria. NIHSS 5. His condition was stable until 2021 when he had symptoms of altered mental state and confusion along with a central lesion of the left facial nerve. A brain MRI showed multiple acute ischemic lesions in the right capsula interna and chronic ischemic lesions in the corona radiata, in the periventricular region and in the cerebellum on the right. Further information form family members was obtained where it was noted that in 1993 the patient had a pilocytic astrocytoma in the region of the hypothalamus. He was treated surgically and also with radiotherapy. He had full remission after treatment however six months later he manifested symptoms of altered mental state, disinhibition, loss of impulse control and hyperphagia. These symptoms resolves spontaneously, however 10 years alter our patients was diagnosed with hypothyroidism and hypocorticism and substitutional hormone therapy was administered. While hospitalized the patients vital signs were stable and his physical examination was normal. He manifested in the neurologic examination disorientation in time and space and residual left sided hemiparesis as well as ataxic gait and dysarthria. NIHSS 5. MMSE 13/30. The laboratory results with biochemistry and coagulation status were all normal. Immunology and multiplate hematologic tests were all normal. Initial brain CT showed an old ischemic lesion frontoparietally on the right and CTA was unremarkable. EEG showed electrocortical dysfunction on the right hemisphere which implies and encephalopathic finding. Holter EKG and transthoracic echocardiography was unsignificant. Neuropsychological examination showed a mixed sensomotor dysphasia and dysarthria as well as moderate cognitive decline with a decrease in all modalities of attention, a severe decline in the verbal declarative thinking process as well verbal declarative memory. Visuoperceptive and visuoconstructive functions as well as executive cognitive functions were also altered. Brain MRI showed old ischemic lesions frontotemporoparietally on the right and periventricular leukoaraiosis and multiple lacunar chronic ischemic lesions supra- and infratentorial. A diagnosis of post-radiation cerebral necrosis was obtained and therapy was initiated. He underwent physical and cognitive and occupational-behavioral. On hospital discharge he was calm and alert, oriented in time and space. The patient was discharged with Kvetiapin tbl 25mg 2x1, Aspirin tbl 100mg 1x1, Atorvastatin tbl 40mg 1x1, physical and cognitive and occupational therapy.

VASCULITIS AS A PRESENTATION OF NEUROBORRELIOSIS

Mirjana Ždraljević[1], Milica Vukojević[1], Marta Jeremić[1], Milija Mijajlović[1,2]

[1]Neurology Clinic, University Clinical Center of Serbia, Belgrade, Serbia
[2]Faculty of Medicine, University of Belgrade, Belgrade, Serbia

Introduction: Neuroborreliosis is an infectious disease caused by the bacterium Borrelia burgdorferi that is transmitted by ticks of the Ixodes genus and occurs in approximately 15% of Lyme disease cases. The neurological disorders caused by this infection include lymphocytic meningoradiculitis with or without paresis, meningoencephalitis, encephalomyelitis, and optic neuritis. Neurovascular disorders are very rare, usually attributed to arteriosclerosis or cerebral embolism. Vasculitis, especially with recurrent strokes, is a very rare manifestation of this disease with estimated frequency between 0.3-1%.

Case presentation: A 54-year-old male was admitted to the Neurology Clinic of the University Clinical Center of Serbia with slow-progressive gait disturbances and cognitive problems. In his personal history, he was treating hypertension and benign prostatic hyperplasia, and had suffered a stroke, which he did not provide documentation of. The first signs of altered walk began more than a year and a half before hospitalization, while six months before hospitalization he began to notice problems in the cognitive domain such as forgetfulness. A detailed neurological examination revealed mild right-sided hemiparesis with distal hypotrophies on the upper and lower extremities. A brain computerized tomography (CT) showed chronic ischemic lesions. On the second day of hospitalization, the patient's existing neurological deficit deepened to the level of severe right-sided hemiparesis and the appearance of new symptoms in the form of speech disorder by the type of dysarthria. Brain CT scan showed no signs of acute ischemia or hemorrhage, while CT angiography of blood vessels of the head and neck revealed stenosis of the M1 segment of the left MCA Intravenous thrombolysis was not administered due to the unknown time of symptoms onset. Magnetic resonance imaging of the endocranium could not be performed due to contraindications, but digital subtraction angiography showed significant stenosis of M1 segment of left MCA and three belonging branches which could be indicative for changes that occur in vasculitis. As part of the extensive diagnostic procedure, a lumbar puncture was performed and the cytobiochemical analysis of the cerebrospinal fluid revealed proteinorachia with a borderline number of cells (Ly 5, PMN 1). CSF sediment for any unusual cells and IgG antibodies against Borelia burgdorferi came back positive. To further investigate this finding, we did line blot test that showed borderline positive IgM antibodies and positive IgG antibodies, so we pursue investigation to a golden standard test CST index of intrathecal synthesis of antibodies that showed us positive results for both IgM and IgG class of antibodies. We did extensive work-up to rule out other diagnoses. After all additional examinations we concluded that our patient probably has vasculitis caused by neuroborreliosis. He was treated with antibiotic (Ceftriaxone 2g/daily during 21 day), antiplatelets (first acetylsalicylic acid, but after a lack of response in the platelet inhibition test, the patient was switched to clopidogrel) and corticosteroid therapy. All these therapeutic measures did not significantly improve the patient's condition, but prevented the occurrence of new ischemic events during follow-up.

Conclusion: Vasculitis is a very rare manifestation of neuroborreliosis. Correct and timely diagnosis is imperative, considering that curing the infection is a very effective way of treating vasculitis.

CASE REPORT- STROKE PFO WITH INITIAL NEGATIVE DWI ON MRI

Juraj Mark Poje

Department of neurology, GH Koprivnica, Croatia

Case presentation: a young female, 37 years old, was admitted to the emergency department of GH Koprivnica with symptoms of diplopia, discrete left hemiparesis and ptosis of the right eye, NIHSS 4, no comorbidities, mRS 0. The symptoms were in regression during the emergency evaluation. Emergency brain CT and angiography were without acute pathological findings, the left vertebral artery is hypoplastic and ends with PICA without signs of occlusion. Emergency MRI is also without pathological findings, without DWI restrictions. The patient presented during the therapeutic window and since the symptoms were in almost complete regression, we also did a TCD monitornig for MES without finding them. We did not decide on an immediate preperfusion treatment due to the resolution of the symptoms. During the next 24 hours, the symptoms are in complete regression. Follow-up MRI after 24 hours shows DWI restriction for acute stroke in the left thalamus. Further extensive evaluation was done with coagulation tests which were all negative, evaluation of risk factors including diabetes mellitus, hyperlipidemia and hypertension and TTE which was without pathological findings. TEE showed an aneurysmal atrial septum with a 13 mm PFO. The patient underwent PFO occlusion with an Aplatzer occluder. A control TCD "bubble" test after 6 months showed 4 MES showing almost complete closure of the PFO. The patient has no symptoms and is on constant antiplatelet therapy.

CAROTID ARTERY DISSECTION AFTER ANABOLIC STEROID ABUSE IN YOUNG PATIENT

Sebastian Andone

Emergency county clinical hospital Targu Mures, Romania

Case presentation: A 33-year-old male patient was found by the family fallen on the floor with right-side motor deficit and speech and understanding difficulties. The patient had no known risk factors (no smoking or previous illness). The patient was found at approximately 13:00, but last time, he was seen without any neurological deficits at 09:00. He arrived at the first hospital (15:22), and the long time was caused by the relative that found him, which led her to think he was drunk.The neurological examination revealed a right central facial palsy, right side hemiplegia (0/5 MRC), right side sensory deficit, and severe mixed aphasia (complete motor aphasia), summing up to 17 points in NIHSS, but 15 points in GCS. The CT scan revealed no hemorrhagic lesions with an ASPECT score of 8 points. The angio-CT scan shows an occlusion of the left internal carotid artery (ICA) after the origin and a secondary occlusion at the M1 segment of the left middle cerebral artery (MCA). The patient was transferred to our center for endovascular treatment, arriving at 18:18. The procedure started at 19:15, with complete recanalization and stent implantation on the left ICA, and the procedure ended at 20:00. Immediately after the procedure, the patient had a minor neurological improvement, only the lower limb from 0/5 to 2/5 MRC; the rest remained the same. The next day, we performed a control CT scan, which revealed a small hematoma (initially suspected to be contrast fluid), but the following days still showed the hyperdense lesion, confirming the

presence of blood. During the hospitalization, we performed a range of paraclinical investigations (CBC, biochemistry, lipid profile, autoimmune panel—ANA panel, antifosfolipid antibodies, infectious panel—hepatitis, syphilis, HIV, thrombophilia panel, cardiac assessment, cervical doppler ultrasound), all of which were in the normal range without any pathological findings. No risk factors were discovered in the history of the patient, nor trauma or heavy exercises were performed in the week before the stroke, although the patient complained of headache one day prior to the stroke. However, after a couple of days, the girlfriend admitted that the patient was taking some tablets for gym purposes without knowledge of the substance contained in the tablets. After she brought the tablets for inspection, we found that the patient was taking anabolic steroids (testosterone-based), and later, after he recovered speech, the patient admitted he had taken a high dose in the last couple of weeks. The clinical evolution was good. The CT scans revealed almost complete blood resorption, and the neurological exam at discharge was greatly improved to a right-side hemiparesis with 1/5 MRC for the upper limb and 4/5 in the lower limb, a minor-moderate motor aphasia with a total NIHSS score of 8 points, and a modified Rankin scale of 3 points. The particularity of the case consists of this being a young patient with no stroke risk factors other than anabolic steroid abuse. Even if the onset time was unknown and we had an extended treatment window, the clinical evolution was good. Although arterial dissections are a common cause of stroke in young adults, dissection due to anabolic steroid abuse is a rare etiology.

MULTIPLE STROKES AND PANCYTOPENIA? DIAGNOSIS IN THE URINE!

Nella Maruskin Gabriel

Neurology Clinic, Dortmund, Germany

Case report: We report on a 56-year-old patient who was transferred to our neurological intensive care unit (ICU) from a surgical department following open reduction and osteosynthesis for multiple new cerebral strokes. Additionally, the patient developed pancytopenia, vasospasms, recurrent multiple strokes, as well as intestinal and splenic ischemia, recurrent sepsis, opportunistic infections, acute renal failure, ultimately leading to multiorgan failure and death. The diagnosis is paroxysmal nocturnal haemoglobinuria. Course: Following a fall resulting in a comminuted per- and subtrochanteric femur fracture on the right side, the patient underwent uncomplicated open reduction and osteosynthesis using a gamma nail on June 5, 2022. Postoperatively, the patient was promptly mobilized and transferred to a regular ward. A few days later, the patient developed left-sided hemiparesis and sepsis of unclear origin with a CRP level of 382 mg/l, necessitating transfer to the ICU and catecholamine support. MRI revealed fresh ischemic lesions in the right medial striate region and scattered punctate ischemic lesions in the left frontal area. CT angiography identified a right M1 stenosis (in duplex sonography bilateral), severe stenosis of the right internal carotid artery (ICA) in the cavernous sinus, additional severe stenosis of the origin of the right ICA and left anterior cerebral artery (ACA) stenosis. With laboratory-confirmed pancytopenia, the patient was placed in reverse isolation. An extensive search for the focus of infection was conducted, yielding no significant findings except for the detection of Candida in the urine, which was considered a possible contamination. After consultation with oncology, the patient received G-CSF filgrastim 30 million IU once daily for pancytopenia, likely associated with

sepsis. Despite a 5-day course of antibiotic treatment with meropenem and vancomycin, the patient experienced a recurrence of fever, accompanied by a rise in CRP level. Clinically, there was a significant deterioration in the patient's condition - she became soporous, progressing to a coma, with oral candidiasis, cutaneous bleeding, a distended abdomen with sluggish peristalsis, tenderness in the lower abdomen and formal tetraplegia. A CT scan of her brain revealed a possible new cortical hypodensity in the right precentral region. A MRI of her brain demonstrated diffuse acute infarcts bilaterally in the frontal regions of the medial and anterior circulation, both cortical and subcortical. Angiographically, there were hypoplastic anterior and middle cerebral arteries bilaterally. Duplex sonography described pseudovenous flows in the M1-M2 segments bilaterally, with partial retrograde flow on the right side. Additionally, a P1-P2 stenosis on the right was noted. Given the presence of progressive cerebral ischemia suggesting vasculitis, Methylprednisolone 1g was administered. Subsequently, due to the detection of further fresh ischemic lesions on CT scans and fluctuating stenoses on duplex sonography, therapy with nimodipine was initiated. During the search for the focus of infection, a CT scan of the thorax revealed a possible atypical infiltrate with a right-sided predominance. No pathogen could be detected at this time. At this point, the patient was already receiving aciclovir, moxifloxacin, voriconazole, vancomycin, caspofungin, and meropenem. Regarding the pancytopenia, a bone marrow aspiration did not reveal an explanatory cause; flow cytometry of the bone marrow aspirate predominantly showed cytolysis. The histopathological findings indicated a significant left shift in myelopoiesis, blast proliferation, massive iron overload, hypoplasia of erythropoiesis and megakaryopoiesis, which would have been unusual to describe as reactive in the context of sepsis. Overall, the patient received 12 units of packed red blood cells and 5 units of platelet concentrates. With abdominal symptoms and increasing lactate levels, we performed a CT scan of the abdomen, which revealed narrowed mesenteric arteries, demarcated splenic infarction ventrally, and a suspected ischemia of the ascending colon to the proximal third of the transverse colon with retroperitoneal perforation. An immediate open right hemicolectomy was performed. Over the next few days, the patient additionally developed acute renal failure, which, after consultation with nephrology colleagues, was describes most likely of multifactorial origin. The elevation of β-D-glucan > 523 pg/ml (normal range < 60 pg/ml) and bronchoalveolar lavage revealed a Pneumocystis jirovecii infection. Unfortunately, two days later, the patient's condition progressed to sepsis-induced multiorgan failure, ultimately resulting in electromechanical dissociation as the cause of death. Diagnosis: The majority of symptoms suggest an explanatory diagnosis: the multiple emboli (multiple strokes, intestinal ischemia, splenic ischemia, microemboli), pancytopenia, opportunistic infections (Pneumocystis jirovecii, candidiasis), vasospasms, renal failure, as well as repeatedly observed laboratory findings such as haemolytic blood picture, elevated LDH, haemoglobinuria, anamnestic complaints of fatigue, miscarriages, post-op exacerbation - all of these explains the diagnosis of paroxysmal nocturnal haemoglobinuria. Paroxysmal nocturnal hemoglobinuria (PNH), also known as Marchiafava-Micheli syndrome, is a rare, potentially life-threatening disorder of hematopoietic stem cells in the bone marrow with variable clinical manifestations. The incidence is approximately 1-1.5:1,000,000/year, but may be underdiagnosed. Prevalence ranges up to 16 cases per million. Onset is most common between the ages of 25 and 45, with women being slightly more affected. Etiologically, it is an acquired mutation in the PIG-A gene (mosaic) in multipotent hematopoietic stem cells resulting in decreased production of glycolipid GPI = protein anchor. This anchors proteins to the cell membrane, especially CD55 and CD59 (known as 'Decay accelerating factor DAF' and 'Membrane-Inhibitor of reactive lysis

MIRL'), which protect cells from attack by the complement system. This leads to impaired membrane stability, and upon complement activation, erythrocytes become sensitive to terminal complement-mediated lysis. Clinically, the presentation is highly variable. Although the name suggests nocturnal haemoglobinuria, this only affects 25% of patients, most likely due to hypoventilation during sleep, resulting in temporary acidosis and haemolysis. Nevertheless, haemoglobinuria, along with pancytopenia and thrombophilia, constitutes the classical triad. Haemolysis leads to endothelial dysfunction due to intravascular release of haemoglobin. Increased metabolism of haemoglobin to methaemoglobin and release of erythrocyte arginase, which reduces arginine as a substrate for de novo NO production, results in vasospasms and platelet activation. Thrombophilia then leads to multiple thromboembolisms - venous and arterial, in typical as well as atypical locations (e.g. myocardial infarction, stroke, abdominal, especially splanchnic vessels, retinal, cerebral veins, cutaneous veins etc.). Thromboembolic complications are the most clinically relevant and are the main cause of increased morbidity and mortality in PNH. The frequently observed renal failure can be explained by vasospasm of the afferent vessels, hemosiderin deposition in the proximal tubules and renal parenchymal damage with impaired tubular function due to microvascular thrombosis. Cytopenia can range from mild, subclinical cytopenia to severe pancytopenia and logically leads to a predisposition to opportunistic infections. Additionally, intermittent oesophageal spasms, chest pain, nausea and swallowing difficulties occur, along with erectile dysfunction in men and miscarriages in women. Furthermore, back pain, headaches, muscle pain, and abdominal pain occur in association with haemolytic crises. The crises/onset of PNH can be triggered by intense physical exertion, infections, surgeries, but also by contrast agent administration. The gold standard for diagnosis is flow cytometry, which is always indicated for thromboembolism (regardless of its location) in association with unclear cytopenia. In terms of therapy, there has been significant progress in recent years. In 1990, half of the patients died within 10 years of diagnosis. Until 2007 PNH could only be treated symptomatically (e.g. blood transfusions), with severe cases undergoing bone marrow transplantation. Since 2007 therapy with Eculizumab has been approved - a monoclonal antibody against complement factor C5. Under this treatment, patients have the same life expectancy as healthy individuals. Most recently in 2019, Ravulizumab and in 2021 Pegcetacoplan were approved.

Conclusion: This case report demonstrates that a diverse array of symptoms should not only be attributed to complications of various conditions, but also that relatively rare diagnosis explaining multiple symptoms should be considered. Nevertheless, in cases of multiple thromboembolic events and pancytopenia, PNH should be always considered!

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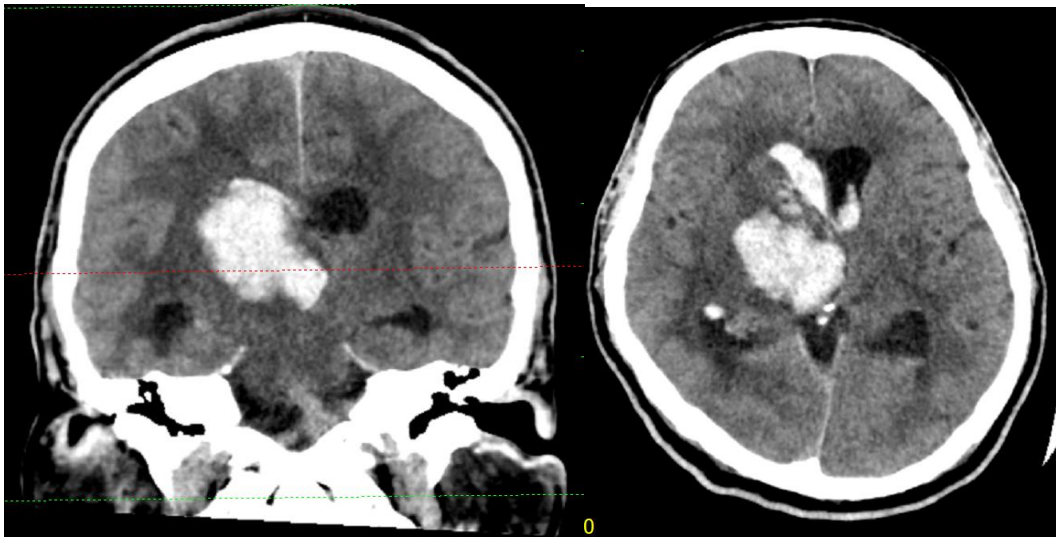
CASE REPORT: RARE CAUSES OF STROKE FROM EVERYDAY CLINICAL PRACTICE
Potential association between a mutation in COL4A5 and brain manifestation

Kateřina Benešová
Department of Neurology, Second Faculty of Medicine, Charles University and Motol University Hospital in Prague, Czech Republic

Introduction: Cerebral small vessel disease (cSVD) is a broad spectrum of diseases affecting the small cerebral vessels and is a significant cause of stroke. While most cases of cSVD are caused by multifactorial brain damage resulting from hypertension, diabetes, or the accumulation of ß-amyloid, a minority of cases have a monogenic cause. Various genes have been reported to cause cSVD with varying incidence in the population. The purpose of this case report is to present a patient with a pathogenic mutation in the *COL4A5* gene, which is associated with Alport syndrome and has yet not been confirmed as a cause of small vessel disease.

Case presentation: The patient was first seen at our Department in April 2023. He was a 66-year-old man with a medical history of untreated arterial hypertension, renal cysts verified by an ultrasound, and benign prostatic hyperplasia. He was on oral therapy with Tamsulosin 0.4 mg per day. There was no history of neurological, cardiovascular, or autoimmune diseases that would be known to run in his family. The patient was a non-smoker and non-alcohol drinker who worked as a bookseller and lived with his wife and two children. The patient was found unconscious on the couch by his son and was immediately transported to the hospital by paramedics. Upon arrival, the patient was assessed with a Glasgow Coma Scale (GCS) score of 3, requiring immediate intubation and mechanical ventilation. In the hospital, a brain CT scan was performed, and it revealed a massive intracerebral haemorrhage in the basal ganglia, with blood extension to the ventricles and a midline shift of 12 mm on the left side, resulting in an ICH score of 4. Therefore an external ventricular drainage was performed by neurosurgeons.

Image NO.1+2: Brain CT scan upon admission to the hospital, coronal and axial scan.



Upon admission to the Neurointensive Care Unit, the patient's GCS score remained the same due to analgosedation. The patient suffered from persistent hypertension, requiring continuous intravenous correction by Urapidil, resulting in stabilisation of the blood pressure around 152/90 torr. The patient also exhibited an irregular heart rhythm with atrial fibrillation. Therefore, a low dose of LMWH and a beta blocker were administered after a control brain CT. Antiedema therapy with mannitol was initiated, followed by a reduction of analgosedation. However, the patient was unable to regain consciousness or breathe without mechanical support, necessitating a tracheostomy. Subsequent brain CT scans did not reveal any further bleeding progression. The patient's stay at the Neurointensive Care Unit was complicated by MDR Pseudomonas aeruginosa sepsis, which led to hemodynamic instability and acute renal failure, requiring the patient to be transferred to the Critical Care Resuscitation Unit. There a continuous venovenous hemodialysis and intravenous antibiotic therapy was initiated, followed by intermittent hemodialysis upon partial resolution of diuresis. The patient's critical condition was further complicated by the development of metabolic acidosis. Looking back, the acute kidney injury and acute tubular necrosis were caused by a combination of factors, including the CT contrast, mannitol intake, and sepsis. After stabilisation, the patient was readmitted to the Neurointensive Care Unit. At the time, he was in a vegetative state with a GCS score of 7, tetraplegia with left-sided spasticity, and critical illness neuromyopathy. A PEG insertion was performed without any major complications. In June 2023, the patient was transferred to the Critical Care Follow-up Clinic. The MRI of the brain was conducted in June before the transfer. The results showed partially resorbed intracranial haemorrhage and improved midline shift to the left. Additionally, there were significant periventricular white matter hyperintensities (Fazekas grade 3) with multiple microbleeds in basal ganglia and cortical locations.

Image NO. 3+4 : Brain MRI, 1 month after ICH: FLAIR sequence, axial scan

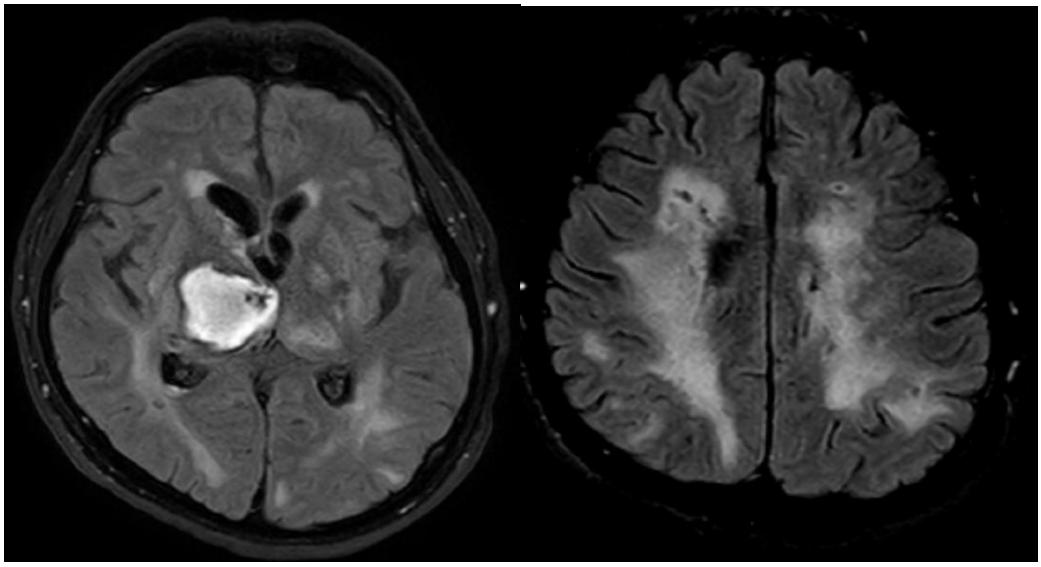
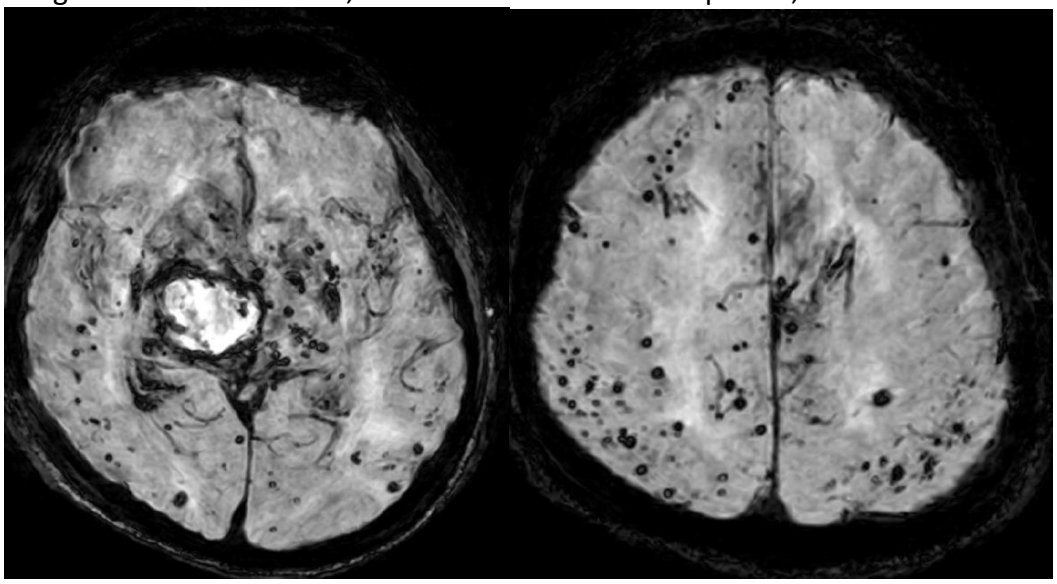


Image NO. 5+6: Brain MRI, 1 month after ICH: SWI sequence, axial scan



Based on the MRI findings, which did not correspond well with the patient's vascular risk factors (only arterial hypertension of unknown level and persistence), as well as his age and medical history, we considered the possibility of conducting a genetic examination of the patient and his family. In 2023, together with the Department of Clinical Genetics, we established a Neurovascular Gene Panel, which covers many genes associated with monogenic brain and vascular conditions, including disease such as CADASIL, various collagenopathies or brain cavernomas. After obtaining informed consent from the patient's children, Next Generation Sequencing was performed. The results revealed a pathogenic variant 1871G>A, p.(Gly624Asp) in the *COL4A5* gene. The patient was found to be hemizygous for this variant. The *COL4A5* gene is associated with Alport syndrome, a genetic condition that causes progressive renal dysfunction and extrarenal symptoms such as hearing impairment, eye abnormalities, and aortic aneurysms. The disease is inherited in 80% of cases in an X-linked pattern. The severity of the disease varies depending on the specific gene variant. Most variants present with hearing impairment in mid-twenties, and by age the of 30, 50% of the males require dialysis or kidney transplantation. However, this exact pathogenic variant is described as a mild form of the disease, requiring dialysis only in 10% of patients. At the time,

the patient's daughter was found to be a heterozygous carrier of the same mutation, without any clinical symptoms. The patient's son tested negative for the mutation.
Discussion: The neurological manifestation of Alport syndrome has yet not been described. One case report has described a possible association between a missense mutation in COL4A5 and cerebrovascular fibromuscular dysplasia. However, intracranial aneurysms are poorly studied and reported in the literature, although there is a clear association between mutations in COL4A5 and aortic aneurysm formation. From a pathophysiological point of view, this gene is responsible for collagen formation, so its dysfunction could lead to collagenopathy, which is a risk factor for physiological blood vessel formation. However, in this case, it is difficult to assess a clear outcome due to the patient's hypertension of unknown duration and levels, which may have caused a small vessel disease. It is important to note that the patient's medical history was obtained solely from family members and not from the patient himself. This could potentially impact the accuracy of the medical history.
Conclusion: Our aim was to describe a potential association between a mutation in COL4A5 and brain manifestation that has not been previously reported. However, further research is needed to establish a definitive association. It is also important to conduct follow-up examinations of family members.

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NON-ATHEROSCLEROTIC INTRACRANIAL STENOSES - A LIKELY MISDIAGNOSED CAUSE OF ISCHAEMIC STROKE IN YOUNG PATIENTS

Marina Krasteva
Clinic of Neurology, University hospital ,,Queen Giovanna – ISUL", Sofia, Bulgaria

Introduction: Non-atherosclerotic intracranial stenoses are a heterogenous group, representing multiple distinct diseases with different etiology, pathophysiology, clinical course, therapeutic approach and prognosis. We present a case report of a young female with ischaemic stroke and bilateral severe non-atherosclerotic intracranial stenoses. A long-term

clinical, neuroimaging and neurosonologic follow-up was performed, demonstrating evolution and prognosis of intracranial stenoses.

Case presentation: A 31-year-old female patient with neither significant previous medical history nor known vascular risk factors suddenly experienced right-sided weakness and sensory disturbance. On emergency admission, 93 minutes after symptom onset, neurological examination revealed right-sided hemiparesis, facial and hypoglossal nerve palsy, as well as right-sided hemihypesthesia (NIHSS=9). Other physical examination detected no abnormalities, and laboratory investigations and electrocardiogram (ECG) were normal. Cerebral computer tomography (CT) demonstrated an acute ischemic lesion in the territory of the left middle cerebral artery (MCA). A color-coded duplex sonography showed bilateral stenoses of the MCA with peak systolic velocity of the the left M1-segment of MCA - 350 cm/sec and right M1 – 230 cm/sec. A conservative therapy with aspirin 100mg/d and atorvastatin 40mg/d was initiated. Cerebral magnetic resonance imaging (MRI) and angiography (MRA) performed 72 hours after symptom onset confirmed the ischaemic lesion in the territory of the left MCA, as well as the bilateral MCA stenoses. Our patient was discharged home on conservative therapy and her neurological symptoms resolved completely within approximately 3 months. During her follow-up, 5 and 7 years later, head MRI didn't reveal any new ischaemic lesions and color-coded duplex sonography confirmed bilateral MCA stenoses without any progression. Our patient was fully asymptomatic on long-term antiplatelet therapy with aspirin 100 mg/d.

Conclusion: Based on the young age of our previous healthy female patient, the bilateral occurence of severe stenoses of the distal ICA, as well as the lack of progression during the follow-up, a final diagnosis of fibromuscular dysplasia (FMD) was made.

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