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Case Report

Isolated arteritis misdiagnosed as bilateral orbital tumors in a patient with acute ischemic stroke ☆,☆☆

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ABSTRACT

Vasculitis is a heterogeneous group of disorders characterized by multifocal segmental inflammation of the small and medium vessels of the central nervous system. The predominant symptoms of cerebral vasculitis are stroke, headache, and encephalopathy. Additional symptoms include seizures, cranial nerve palsies, and myelopathy. Imaging techniques play a crucial role in identifying the diagnosis of vasculitis and demonstrating brain involvement. An 89-year-old woman with permanent atrial fibrillation developed an embolic stroke. In treatment, intravenous thrombolysis and thrombectomy with complete antegrade reperfusion of the left middle cerebral artery was used, without the clinical effectiveness. Brain MRI revealed bilateral oval lesions in medial parts of the orbits, which were initially misinterpreted as orbital tumors. Final diagnosis confirmed thickened arterial walls as orbital changes due to inflammatory arteritis. Ten days later, follow-up MRI was performed and showed complete regression of the orbital masses. Primary central nervous system vasculitis, manifesting as acute ischemic stroke, may be reversible with early systemic thrombolytic treatment.

Abbreviations: ACA, anterior cerebral artery; BRAO, branch retinal artery occlusion; CNS, central nervous system; CRAO, central retinal artery; CSF, cerebrospinal fluid; CTA, CT angiography; VEGF, endothelial growth factor; EGPA, eosinophilic granulomatosis with polyangiitis; GCA, giant cell arteritis; GPA, granulomatosis with polyangiitis; ICA, internal carotid artery; LVV, large vessel vasculitis; MPA, microscopic polyangiitis; MCA, middle cerebral artery; PDGF, platelet-derived growth factor; PACNS, primary angiitis of the central nervous system; RAO, retinal artery occlusion.

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Introduction

Cerebral vasculitis is a term that tends to describe inflammation in the blood vessel wall of the central nervous system (CNS), associated with pathological changes, occlusion and infarction. In “primary” (also called “isolated”) CNS vasculitis, there is little or no generalized inflammation. “Secondary” CNS vasculitis occurs when the CNS becomes involved in systemic vascular disease, including, but not limited to, systemic vascular diseases such as microscopic arteritis or granulomatosis with vasculitis. All forms of cerebral vasculitis are relatively rare, but all are serious and potentially life-threatening [1,2].

Case report

An 89-year-old woman with persistent atrial fibrillation (without anticoagulation) and hypertension was admitted to Department of Neurology with a speech problem and right-side weakness of the limbs. On neurological examination at the arrival, she was conscious, with global aphasia, right asymmetry of the face, paresis of the right limbs with lower tone and right side Babinski sign. She had 19 points in the NIHSS scale.

The patient had no history of COVID-19 disease. During hospitalization, there were no symptoms suggestive of SARS-CoV-2 infection. On hospital admission and on the third day of admission, negative results were obtained from genetic testing of nasopharyngeal swab material for coronavirus 2 (SARS-CoV-2) infection by real-time molecular polymerase chain reaction (RT-PCR). The patient had a history of being vaccinated with two doses of mRNA vaccine against COVID-19 (last dose taken 2 months before the stroke).

Initial noncontrast CT revealed early ischemic changes in the left hemisphere in the area of the insula and frontal lobe. CT angiography (CTA) showed acute occlusion of the left MCA in part M1/M2 as well as the chronic occlusion of the left internal carotid artery (ICA) (Fig. 1). Additionally, a mass within the left orbit was reported as an incidental finding on CT (Fig. 2). The patient was qualified to intravenous thrombolysis with improvement and reperfusion left ICA. Conventional angiography showed occlusion of left middle cerebral artery (MCA) and normal blood flow in left ICA and left anterior cerebral artery (ACA). The mechanical thrombectomy was connected with complete antegrade reperfusion of the left MCA but without clinical effectiveness. A follow-up CT detected an acute ischemic stroke in the area of left MCA. Brain MRI revealed bilateral oval lesions in the medial part of the orbits, which were initially misinterpreted as orbital tumors (Fig. 3). Angio-CT of the chest showed embolic material in the segmental arteries of the lower lobe of the right lung. The echocardiography showed concentric wall hypertrophy, especially of the interventricular septum, degenerative changes in the mitral and aortic valve, tight, low gradient aortic stenosis with preserved left ventricular ejection fraction. Twenty-four-hour Holter monitoring heart rate revealed atrial fibrillation. The 24-hour blood pressure Holter was normal. Ophthalmological examination did not reveal any signs of an embolism of the central retinal artery. She was consulted by an angiologist and a maxillofacial surgeon. Laboratory investigations showed mild raised inflammatory markers: CRP 10.74 mg/dL (reference value $\leq$ 0.5 mg/dL) and procalcitonin 0.11 ng/ml (reference value $\leq$ 0.05 ng/mL). Renal, liver function, and electrolytes were within normal reference ranges. CSF examination reveals mild pleocytosis (10 cells/uL) with mild elevation of proteins (55 mg/dL) and normal glucose. Multiple blood and urine cultures were negative. SARS-CoV-2 infection was also excluded. Further investigations including vasculitic

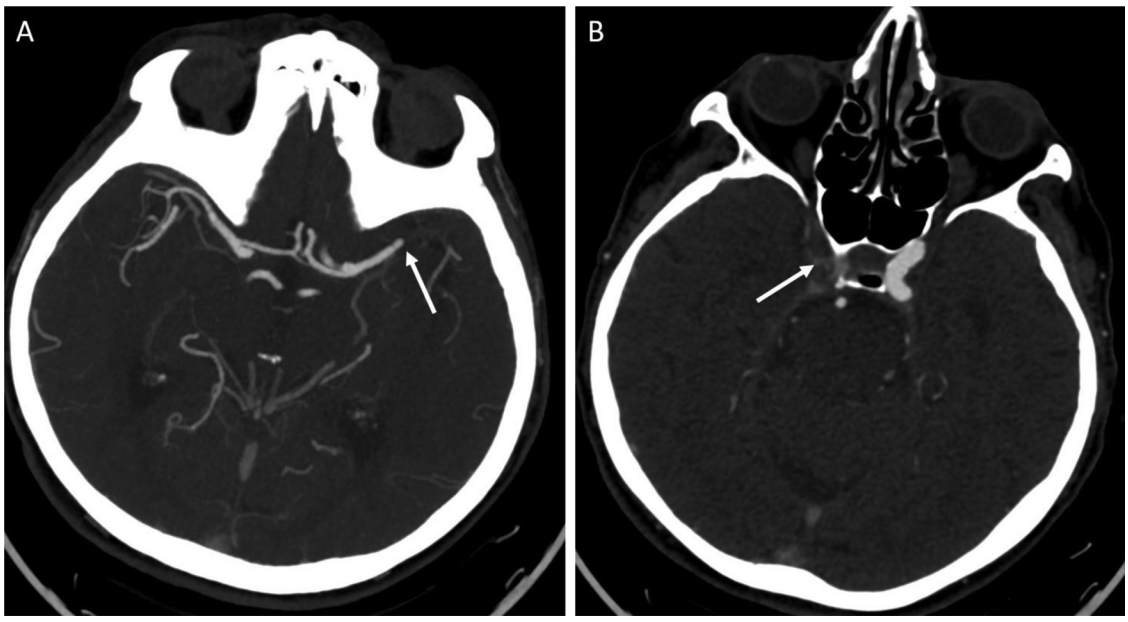

Fig. 1 – CT angiography, axial images (A, B): there is an acute occlusion of the left MCA in part M1/M2 (A) as well as the chronic occlusion of the left internal carotid artery (B) (arrows).

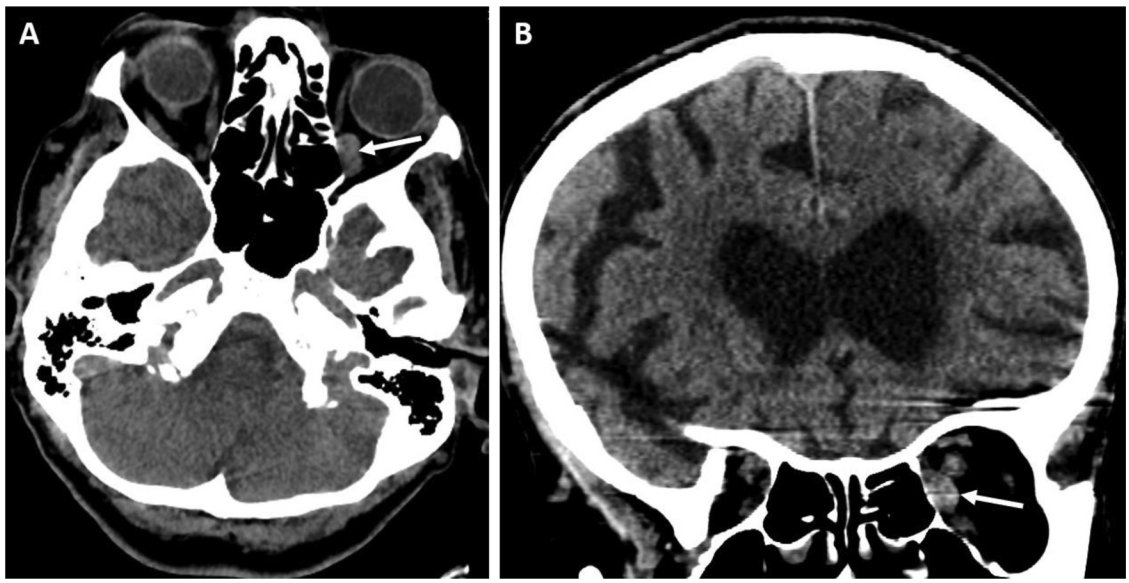

Fig. 2 – CT examination, axial image (A) and coronal reconstruction (B): a mass within the left orbit was reported as an incidental finding on CT (arrow).

screen, complete autoimmune screen, infective viral screen, and malignant screen were either negative or in the normal range. The consulting rheumatologist and angiologist have excluded systemic vasculitis, neoplasm, and histiocytosis. Ten days later, follow-up MRI was performed and revealed an absolute disappearance of the orbital masses (Fig. 4). Final diagnosis was made after a careful reassessment of MRI examination showing the thickened arterial wall due to inflammatory arteritis. The homogenous intense enhancement of the thickened arterial wall on T1-weighted images after contrast administration together with a typical low signal "flow void" inside the vessel seen on T2-weighted images (Fig. 3) as well as a complete regression of changes on the follow-up MRI were the best diagnostic clue.

She was treated with antihypertonic agents, fluid therapy, antibiotics, and diuretics.

In the etiology of stroke in our patient, the authors emphasize the coexistence of primary cerebral vasculitis with associated disseminated embolic process (coexisting pulmonary embolism).

Discussion

Vasculitis is a heterogeneous group of conditions characterized by inflammatory and necrotic changes in the vessel wall. Based on Chapel Hill Consensus Conference criteria (CHCC) criteria, primary systemic vasculitis can be divided into 3 groups: large, medium, and small vessel. Antineutrophil cytoplasmic antibody-associated vasculitis is an important group of small-vessel vasculitis. This group includes granulomatous vasculitis, namely, granulomatosis with polyangiitis (GPA), nongranulomatous vasculitis, namely, microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA) [3–5]. Large-vessel vasculitis, that is, giant cell arteritis (GCA) and Takayasu arteritis, combine autoimmune and autoinflammatory components to an extent depending on acquired or innate immune response.

The authors present a case report in which an 89-year-old female patient with multiple cardiovascular risk factors (hypertension, persistent atrial fibrillation, generalized atherosclerosis) and a history of cerebrovascular lesions, the left MCA occlusion was diagnosed with early cerebral ischemic stroke of the left cerebral hemisphere, treated with reperfusion therapy (intravenous thrombolysis and mechanical thrombectomy). The authors highlight the diagnostic difficulties and distinct therapeutic options. The authors also underline the diagnostic difficulties and the need for an interdisciplinary approach in a patient with vasculitis. Due to complete aphasia, it was impossible to assess the patient's subjective state. The differential diagnosis also considered a retinal artery occlusion (RAO), which may be located in the central retinal artery (central retinal artery occlusion; CRAO) or its branches (branch retinal artery occlusion; BRAO). These emboli may be single or multiple. The clinical picture of RAO depends on the vessel that is occluded by the embolic material. Central retinal artery embolism manifests as a sudden, painless loss of vision, while BRAO may remain asymptomatic or result in only a subtle decrease in visual acuity or sectoral visual field loss. The ophthalmologic diagnosis performed allowed us to exclude RAO. The patient's differential diagnosis included also bilateral orbital tumor, such as intraorbital lymphoma, meningioma, and sarcoidosis, because all these masses demonstrate intense enhancement after contrast administration. The orbital lymphoma typically shows restricted diffusion on DWI and low signal intensity on T2-weighted images. Meningioma presents the characteristic encasement of the optic nerve as this entity is in fact the optic nerve sheath tumor. Apart from that, orbital meningioma usually is a single tumor. Finally, the diagnosis of sarcoidosis is less likely in a case with no history of any respira-

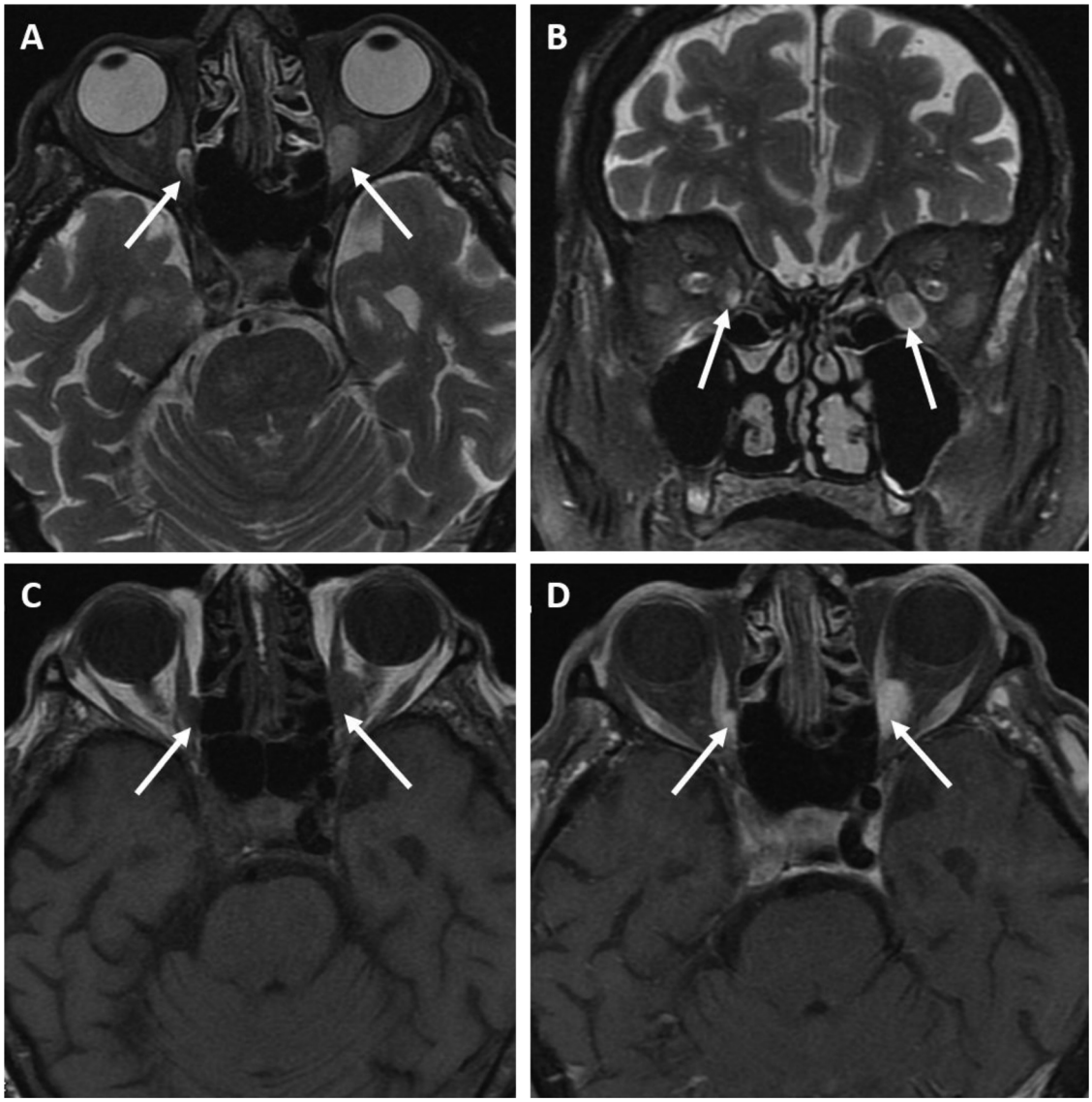

Fig. 3 – MRI examination, axial (A) and coronal T2-weighted FATSAT (B) images, axial T1-weighted unenhanced (C), and enhanced (D) images. MR revealed bilaterally in medial part of the orbits oval lesions (arrows) presenting low signal on T1 and high signal on T2 images. After contrast administration, masses showed intense homogenous enhancement. Additionally, on T2 images (A, B), there was a low signal "flow void" visible inside the oval lesions confirming the nature of the orbital masses to be vessels—these MRI findings were initially not noticed; therefore, the lesions were misinterpreted as orbital tumors.

tory diseases. Moreover, the differential diagnosis of orbital masses includes also common vascular lesions such as capillary (infantile) hemangioma, cavernous hemangioma (solitary encapsulated venous-lymphatic malformation), and lymphangioma (venous-lymphatic malformation) [6]. HighT2 signal, lobular borders, fine internal flow voids, extraconal location, and intense homogeneous enhancement are the typical features of the capillary hemangioma. The key MRI findings of the cavernous hemangioma consist of the intraconal location, a very high T2 signal with hypointense pseudocapsule; and an early nodular enhancement with progressive accumulation of contrast on the later phase images. The lymphangioma is a markedly T2 bright nonenhancing mass with internal septations, with or without layering blood products and solidly enhancing components. Benign tumors, that should also be taken into account, include schwannoma and neurofibroma. Malignancies that should be considered apart from orbital lymphoma are also metastasis as well as rhabdomyosarcomas.

Moreover, IgG4-related diseases should also be considered in the differential diagnosis. A case of a patient with a progressive bilateral tumor of the upper eyelid and orbit has been reported in the literature. The diagnosis of IgG4-related disease was made only after many years and multiple tumor biopsies. Initiation of systemic steroid therapy led to a rapid shrinkage of the tumors and improvement in the patient's overall

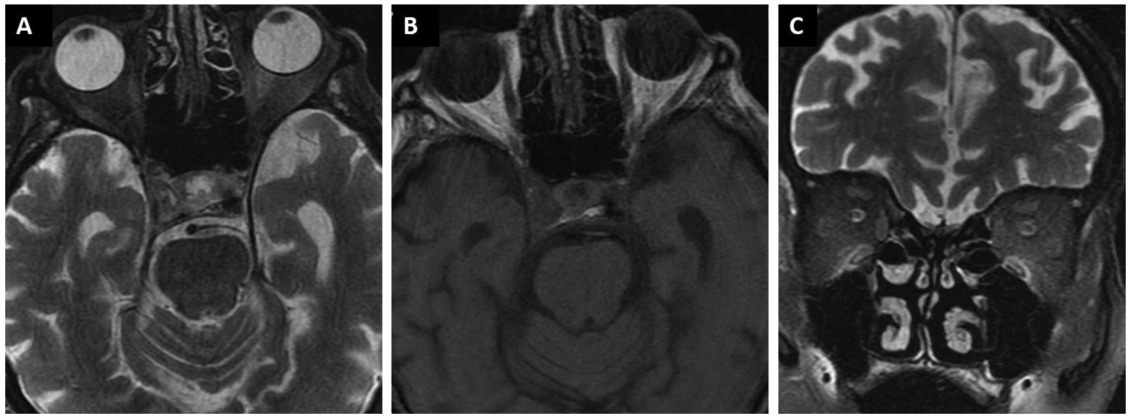

Fig. 4 – Follow-up MRI examination after 10 days, axial T2-weighted FATSAT (A), axial T1-weighted (B), and coronal T2-weighted FATSAT (C) images. The previously seen masses completely disappeared.

condition. IgG4-related disease describes a recent multisystemic disorder that can also be seen in the orbit. Tumor-like mass lesion is a rare presentation of PACNS, accounting for only 5% of all PACNS. Accordingly, it is difficult to differentiate tumor-like PACNS from neoplastic diseases. Jin et al. described a case of PACNS initially misdiagnosed as glioblastoma [7]. Also, Zhang et al. described an extremely rare case of PACNS occurring in the cerebellum, which mimicked a cerebellar tumor [8]. Most PACNS are multiple lesions, occurring in the supraparietal subcortical and deep white matter, and only in a few cases do they occur as a nodular mass lesion. The authors conclude that a possible mechanism of mass lesion formation is the breakdown of the blood-brain barrier in small vessels by inflammatory cell infiltration in perivascular and parenchymal areas, leading to enlargement of the lesion mimicking the mass.

The final diagnosis in the presented case was made after a careful reassessment of the MRI examination. The homogenous intense enhancement of the thickened arterial wall on T1-weighted images after contrast administration together with a typical low signal "flow void" inside the vessel seen on T2-weighted images as well as a complete regression of changes on the follow-up MRI were the best diagnostic clue. The "flow void" relates to a signal loss occurring with blood and other fluids, moving at sufficient velocity relative to the MRI scanner. Identifying flow voids at MRI examination provides valuable information about the flow dynamics or integrity of vascular structures.

A low signal "flow void" visible inside the oval lesions and confirming the nature of the orbital masses to be vessels was initially missed; therefore, the lesions were misinterpreted as orbital tumors.

Conclusions

Thromboembolic complications in vasculitis are an important feature of the disease. The reversible primary angiitis central nervous system vasculitis, simultaneous with embolic stroke, can be the result from early systemic thrombolytic therapy.

The authors highlighted that the differential diagnosis of the enhancing bilateral masses within the orbits should include not only tumors but also thickened arteries due to an inflammatory process. A very useful MRI finding indicating the nature of the lesion is a low signal "flow void" in the central part of a mass seen on T2-weighted images.

Author contributions

Conceptualization: E.D.; writing—original draft preparation: E.D. and JCL.; Radiology: J.B.; Editing J.B. and B.P. All authors have read and agreed to the published version of the manuscript.

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Institutional review board statement

Wrocław Medical University does not require ethical approval for reporting individual cases. All procedures performed involving the human participant were in accordance with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Data availability statement

Not applicable.

Patient consent

The authors certify that they have obtained all appropriate patient consent forms. In this form, the patient has consented to the publication of radiological findings and other clinical information in the journal. The patient understands that their name and initials will not be published and that reasonable efforts will be made to conceal their identity to guarantee anonymity.

Conflicts of Interest

The authors declare no conflict of interest.

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