39. ATYPICAL PRESENTATION OF GIANT CELL ARTERITIS CONFIRMED ON BRAIN BIOPSY

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Introduction: This intriguing case describes a patient in who initial giant cell arteritis (GCA)/temporal arteritis (TA) presentation was preceded by bilateral acute anterior uveitis. He presented several months later after being treated for GCA with new neurological symptoms not typical of ischaemic cerebrovascular accident (CVA) on brain imaging. After ruling out a variety of differentials including an infection, he was treated for cerebral vasculitis secondary to temporal arteritis confirmed on brain biopsy which remains gold standard for diagnosis.

Case description: A 73-year-old patient with a background history of hypertension and mild asthma presented with three week history of ocular pain, headache and photosensitivity after a fall. CT head and lumbar puncture (LP) were unremarkable. He was diagnosed with bilateral acute anterior uveitis by ophthalmologists and treated with topical cyclopentolate and dexamethasone . In view of headaches, scalp tenderness, jaw claudication and raised inflammatory markers he was treated with 60mg of prednisolone for presumed giant cell arteritis (GCA) and temporal artery biopsy (TAB) was organised.

He showed marked symptomatic improvement on steroids. Inflammatory markers normalised (erythrocyte sedimentation rate (ESR) 77 $\rightarrow$ 5 and C-reactive protein (CRP) 130 $\rightarrow$ $<$1). Temporal artery biopsy was negative, but took more than four weeks after starting steroids and was only 9mm in length. Serum screening was unremarkable for complements C3,4, antinuclear antibodies (ANA), anti neutrophil cytoplasmic antibodies (ANCA), bacterial or viral antibodies.

Ten months later he was admitted with a two-week history of gradually worsening bilateral lower limb weakness on the background of chronic lower back pain. Magnetic resonance imaging (MRI) head showed parasagittal abnormalities which were thought to be atypical for ischemic infarction. Intracranial angiogram did not reveal any pathology. LP demonstrated elevated white cells (18 $\times$ 106/L – normal $<$5 $\times$ 106/L) and protein 0.61g/L (normal $<$ 0.15-0.45g/L) with negative oligoclonal bands. The serology for neuronal autoantibodies and quantiferon was negative. ESR was elevated (50). Echocardiogram showed no vegetations.

He was managed for acute cerebral vasculitis with methylprednisolone and pulsed cyclophosphamide (CYC). He also underwent a repeat TAB which was normal. In view of clinical deterioration he underwent repeat MRI head and spine which showed persistent active inflammation. Brain biopsy was organised which confirmed granulomatous inflammation with multinucleated giant cells. Unfortunately he continued to deteriorate, suffered from multiple infections and sadly passed away at his home with his family.

Discussion: Giant cell arteritis is a systemic vasculitis characterized by granulomatous inflammation of aorta and its main vessels. Visual complications are mostly due to vasculitis of posterior ciliary arteries. Uveitis as a presenting feature of GCA is uncommon. We should be aware that, although unusual, uveitis in elderly patients can be a presenting feature of GCA.

Cardiovascular risk is increased in these patients. Several case series of myocardial infarction and stroke have been reported. About 30% of patients present with neurological manifestations, the most common are neuropathies (14%), including mono- and polyneuropathies of the limbs; stroke has been extensively described (5-20%), particularly vertebrobasilar ischemia.

Cerebral vasculitis may occur as primary angiitis of the central nervous system (PACNS) or as CNS manifestation of systemic vasculitis. In GCA, the involvement of CNS arteries is very rare ($<$2%).

Our patient's imaging revealed bilateral parafalcine frontal lobe changes in anterior cerebral artery territory. However, infarction in this territorial area is quite rare unless there is space occupying lesion or anatomical anomalies of vasculature. In our patient the MRI appearances were not convincing for ischaemic infarction.

Major symptoms of cerebral vasculitis are stroke, headache and encephalopathy. Diagnosis is based on a combination of clinical, laboratory and imaging findings. In systemic vasculitis an acute inflammatory response with raised ESR and CRP may be present. CSF studies reveal mild lymphomonocytic pleocytosis or protein elevation in more than 90%. Magnetic resonance imaging, with or without contrast, is the investigation of choice to detect and monitor cerebral involvement. The treatment recommendations are derived from protocols for systemic vasculitides. A combination of steroids and pulse cyclophosphamide (CYC) is recommended for induction treatment. Methotrexate, azathioprine and mycophenolate mofetil can be used for maintenance therapy similar to ANCA associated vasculitis.

Key learning points: Our case highlighted the rare presenting feature of GCA in the form of bilateral uveitis.

Our patient was at high risk for developing ischaemic cerebral vascular event in view of large vessel vasculitis, his age and co-morbid hypertension but radiological imaging wasn't typical for this and raised the suspicion of active cerebral vasculitis.

One should suspect multifocal brain disease like vasculitis when neurological deficit can't be explained easily by territorial distribution of cerebral circulation. Cerebral vasculitis can be suspected on brain imaging and confirmed with biopsy.

It is important to make this diagnosis as the treatment is immunosuppression different from that of a typical stroke and can be rewarding.

Our patient was managed with immunosuppressive therapy but continued to deteriorate that prompted the need for brain biopsy which remains the gold standard for diagnosing cerebral vasculitis.

Conflicts of interest: The authors have declared no conflicts of interest.