Catastrophic primary central nervous system vasculitis

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ABSTRACT

Primary central nervous system vasculitis (PCNSV) is an uncommon condition that affects the brain and the spinal cord. It is heterogeneous in presenting characteristics and outcomes. We report a patient with a catastrophic rapidly progressive course refractory to intensive treatment with pulses of methylprednisolone and iv cyclophosphamide. The condition rapidly deteriorated and the patient died 6 weeks after presentation. Rapidly progressive PCNSV represents the worst end of the clinical spectrum of PCNSV. These patients are characterised by bilateral, multiple, large cerebral vessel lesions on angiograms and multiple bilateral cerebral infarctions.

A 47-year-old previously healthy man was admitted for a 2-week history of multiple cerebral infarcts. His condition rapidly deteriorated, and he had a right hemiparesis and lethargy. MRI showed multiple infarcts of various ages (Fig. 1A-B). MR and conventional angiograms showed stenoses in multiple cerebral arteries, bilaterally (Fig. 1D-E-F). The patient did not report a history of exposure to vasoactive substances, and did not have thunderclap headaches, or other manifestations typical of reversible cerebral vasoconstriction syndrome. Extensive blood and spinal fluid investigations excluded systemic vasculitis, connective tissue diseases and infections. ESR was 3 mm/hour, spinal fluid showed mildly elevated protein levels

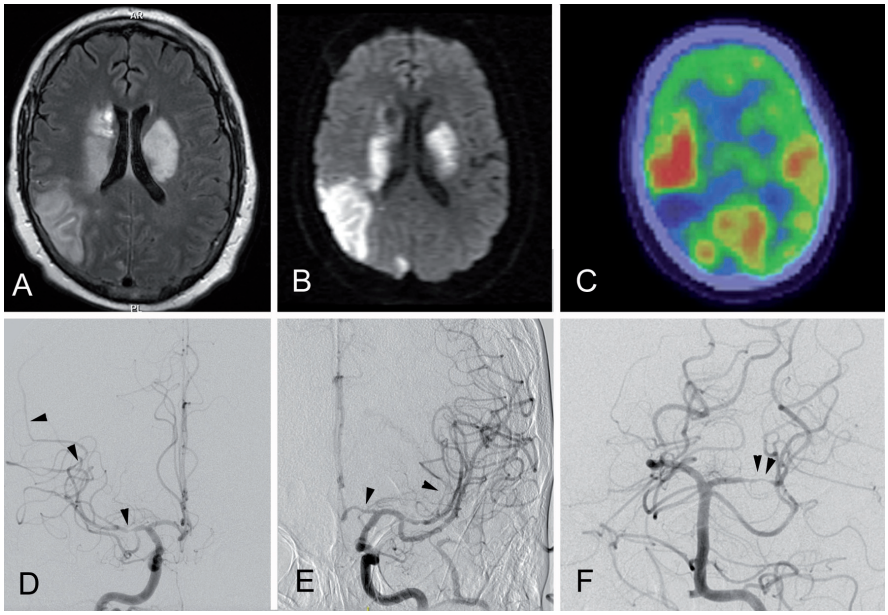

Fig. 1. MRI, FDG-PET, and DSA angiogram of the patient. **A**. Axial Flair Image demonstrating mass like cytotoxic oedema in the right parietal lobe and bilateral deep grey nuclei. **B**. DWI demonstrating acute infarcts (restricted diffusion) in multiple vascular territories including the lenticulostriate arteries bilaterally, right MCA and right PCA territories. **C**. Axial FDG-PET scan demonstrates hypometabolism in the region of acute infarctions and increased FDG accumulation along the anterior edge of the right parietal and occipital infarcts consistent with luxury perfusion. **D**-**E**. Digital subtraction angiography (DSA) from bilateral ICA injections demonstrates multiple regions of alternating narrowing and dilatation (black arrowheads) of the M1 and M2 MCA and left A1 ACA branches consistent with vasculitis. **F**. DSA from a left vertebral injection demonstrates marked irregular narrowing of the P1 segment of the left PCA.

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(69 mg/dL). FDG CT/PET, performed to evaluate for lymphoma that can be associated to cerebral vasculitis (1), was abnormal only in the brain showing hypometabolism in the region of acute infarctions and increased FDG accumulation along the anterior edge of the right parietal and occipital infarcts consistent with luxury perfusion (Fig. 1C). A diagnosis of primary central nervous system vasculitis (PCNSV) was made, and despite the intense treatment with pulses of methylprednisolone and iv cyclophosphamide the condition deteriorated and the patient died one month later.

Catastrophic PCNSV represents the most ominous subset of this vasculitis characterised by several subsets that can differ in terms of prognosis and optimum management (2-5). Often it has a fatal outcome, typically involves the larger cerebral arteries bilaterally with the occurrence of multiple cerebral infarctions, and the response to conventional immunosuppressive treatment is poor.

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