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Central nervous system vasculitis presenting as an ischaemic stroke in a young woman with systemic sclerosis

A 29-year-old woman with diffuse cutaneous SSc for 7 years presented with the acute onset of right hemiparesis. She had a regular pulse of 90/min and blood pressure 126/84 mmHg. Power in the right arm and leg was initially 4/5 but rapidly worsened to 3/5 in the leg with a positive Babinski sign.

A plain CT scan of the head was normal. An MRI (Fig. 1) revealed a large infarct in the territory of the left anterior cerebral artery. An ECG, echocardiogram and Doppler of the extracranial vessels were normal. aPL were absent. The ESR was 87 mm/h. The patient was treated for possible CNS vasculitis using i.v. pulse CYC, with rapid improvement. She recovered completely by the third monthly dose of CYC.

The incidence of stroke was 2.61 times higher in 865 patients with SSc than in controls in a large cohort study [1]. Most strokes in this group were likely to be atherosclerotic. Only a few cases of CNS vasculitis have been reported in patients with SSc [2]. Given the absence of risk factors for atherosclerosis, we conclude that the likely cause for the ischaemic stroke in our patient was CNS vasculitis.

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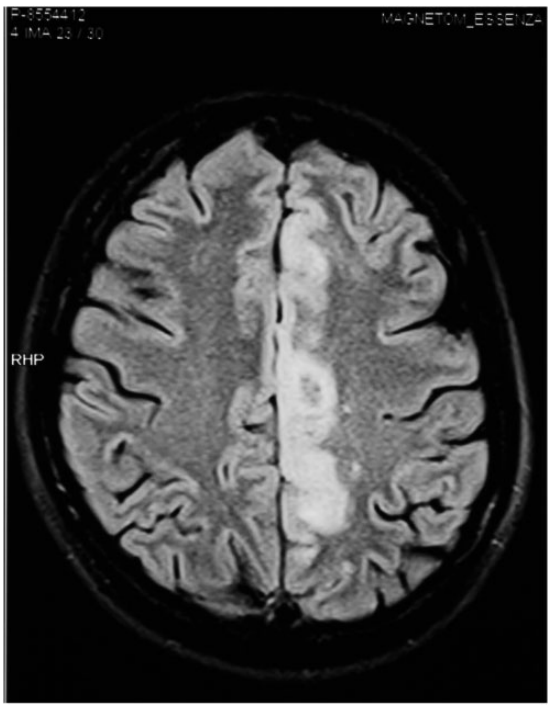
Fig. 1 MRI of brain showing infarct in the left anterior cerebral artery territory



Axial section of fluid-attenuated inversion recovery (FLAIR) sequence of MRI of the brain showing hyperintensity in left parafalcine frontotemporal region, suggesting an acute infarct in the territory of the left anterior cerebral artery.

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