A sleepless young man with red eyes and slurred speech?

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Learning Point for Clinicians

Ischaemic strokes can be the first presentation of ANCA-associated vasculitis, not only in patients with established disease. They can involve small vessels supplying subcortical structures, including the basal ganglia and thalamus. Early recognition and immunosuppression of ANCA-associated vasculitis prevents life-threatening pulmonary and renal complications while likely to improve outcomes from stroke.

A 37-year-old Indian gentleman presented to the emergency department with sudden-onset slurred speech associated with recent sleep disturbances and a 2-year history of recurrent sinusitis, nose bleeds and intermittent redness of his eyes. There was no associated headache, word finding difficulties, focal weakness, visual deficits or cognitive impairment. There was no haemoptysis, shortness of breath and no previous history or risk factors for cardiovascular disease.

Examination revealed bilateral episcleritis and dysarthria in the absence of expressive or receptive dysphasia. There was no focal neurological deficit or extrapyramidal signs and the patient scored fully on the abbreviated mental test. The remainder of the physical examination was unremarkable and urine dipstick was negative for blood and protein.

Diffusion-weighted MRI of the brain revealed acute bilateral basal ganglia ischaemic infarcts in the lateral lenticulostriate territories. A chest radiograph, lumbar puncture, echocardiogram, cardiac 24-hr tape and carotid ultrasonography were unremarkable. The erythrocyte sedimentation rate was 34 mm/hr; C-reactive protein 36 mg/l; white cell count 7.8 with no eosinophilia, serum anti-neutrophil cytoplasmic antibody (ANCA) was strongly positive for proteinase 3 ANCA antibodies (36IU, normal range <3IU). A CT scan of the sinuses revealed extensive maxillary and sphenoid sinusitis. Lung function tests were normal. A diagnosis of ANCA-associated granulomatosis with polyangiitis (GPA, formerly Wegener's granulomatosis) with cerebral vasculitis and basal ganglia infarction was made on the basis of sinusitis, raised inflammatory markers, presence of PR3-ANCA positivity and MRI findings.

The patient was commenced on 300 mg aspirin, subsequently changed to clopidogrel after 14 days. On diagnosis of GPA, the patient was treated with a pulsed regime of intravenous methylprednisolone, followed by high-dose oral prednisolone, and intravenous cyclophosphamide was commenced in accordance with the EUVAS protocol.1 After completion of therapy the patient was asymptomatic with no neurological deficits and normal inflammatory markers. Repeat ANCA testing was negative at 6 months, indicating disease remission, and is currently maintained with 5 mg prednisolone and azathioprine. The patient has returned to work and remains well.

Discussion

This report describes an unusual case of bilateral basal ganglia infarction as a first presentation of...
ANCA-associated GPA (Wegener’s granulomatosis). Although ischemic stroke is well described in large vessel vasculitides, it is a rare manifestation in ANCA-associated vasculitides. The majority of ANCA-associated vasculitic strokes occur after a diagnosis of vasculitis has already been established, when the risk of stroke may be augmented by associated vascular inflammation.2

The thalamus is known to play in important role in sleep regulation and paramedian strokes have previously been associated with hypersomnia, paroxysmal sleep and non-REM sleep irritability.3 The unusual symmetrical thalamic involvement of stroke in our described case was attributed to vasculitic occlusion of a single artery of Percheron. This anatomical variant, found in 10% of the normal population,4 arises from the P1 branch of the posterior cerebral arteries and results in a solitary common thalamic arterial supply to both paramedian regions, instead of two separate thalamic arteries.

ANCA vasculitis must be considered in patients with atypical patterns of stroke that have raised inflammatory markers and lack classic cardiovascular risk factors. Furthermore, involvement of a single, common arterial source such as the artery of Percheron and an underlying diagnosis of ANCA-associated vasculitis must be considered in patients presenting with bilateral, symmetrical lesions in deep small vessel territories. Early recognition of vasculitis is paramount as prompt immunosuppression with steroids and systemic chemotherapy may improve outcomes from stroke and will prevent life-threatening complications of pulmonary and renal vasculitis.5

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References


