Giant cell arteritis presenting as lateral medullary syndrome

Sir, Stroke referable to the vertebrobasilar territory is an uncommon clinical manifestation of giant cell arteritis. It accounts for <2% of the neurological presentations of this condition [1]. This case report describes a case of temporal arteritis presenting as lateral medullary syndrome in an elderly man.

A 74-yr-old man presented to hospital with a 2 day history of sudden onset of ataxia, mild visual blurring, dysarthria and headache. He had a 1 yr history of angina, but no history of stroke, diabetes or hypertension. He took metoprolol 50 mg twice daily and aspirin 300 mg three times a week. There were no symptoms of previous headache, jaw claudication or other systemic features suggestive of arteritis. He smoked a pipe (2 oz of tobacco/week).

On initial examination, his heart rate was 70 and regular, his temperature was 36°C. his respiratory rate was 16 and his blood pressure was 140/70. Neurologically, he was alert and orientated. Examination of his cranial nerves revealed a left
Horner’s syndrome, fine lateral nystagmus to the left and right, a decrease in pinprick sensation over the left face and an absent left corneal reflex. He had a left palatal palsy with dysarthria and he coughed on swallowing sips of water. Examination of his limbs revealed decreased pinprick sensation and temperature on the right leg and trunk to level T2/3. His gait was broad based and he consistently tended to fall to the left. A nasogastric tube was inserted for feeding purposes.

Initial investigations showed a normal ECG, chest X-ray and CT scan of the brain. His Hb was 11.0 g, WBC 10.4, platelet count 485 and ESR 88 mm/h. Serum electrolytes and liver function tests were normal. The following day an MRI was performed (Fig. 1) which confirmed an area of high signal abnormality in the left lateral medulla on the T2-weighted image consistent with a recent infarct. Doppler ultrasound showed anterograde flow in the vertebrobasilar system and no evidence of carotid disease. Transthoracic echocardiography showed no evidence of thrombus.

Despite repeated clinical examination, a septic screen and investigation for occult malignancy, no cause for the raised ESR was found. A temporal artery biopsy was performed. This revealed a panarteritis with an extensive mononuclear cell infiltrate and fragmentation of the internal elastic lamina consistent with giant cell arteritis.

The patient was treated with oral prednisolone, initially at 60 mg/day, and intense rehabilitation for his stroke. His recovery was complicated by a left corneal ulcer. This was treated topically with chloromycetin, but was slow to settle. During the period of the abrasion, his ESR was noted over a week to be increasing on prednisolone 25 mg/day. The prednisolone was therefore increased to 40 mg/day for 5 days, with rapid healing of the abrasion. The remainder of his clinical course was uncomplicated and he made a good recovery. He was able to return home, eating normally and with a mild ataxia only.

Giant cell arteritis is limited to vessels with an internal elastic lamina. There is a close correlation between the susceptibility to arteritis and the amount of elastic tissue present in an arterial wall. As a consequence, the highest incidence of severe involvement in the head and neck is in the superficial temporal arteries, the vertebral arteries, and the ophthalmic and posterior ciliary arteries [2]. The internal carotid, external carotid and central retinal arteries are affected less frequently, and intracranial vessels are rarely involved. Involvement of the aorta and its branches, the abdominal vessels and coronary arteries has been described.

The distribution of vascular involvement helps explain the relative incidence of the neurological complications. The most common are neuro-opthalmological (most importantly blindness), but virtually every type of neurological manifestation is possible, including neuropsychiatric syndromes, peripheral neuropathies and mononeuropathies, cord lesions, neuro-otological syndromes, various pain syndromes, transient ischaemic attacks and stroke [3]. Infarction in the vertebrobasilar territory is relatively uncommon, occurring in only two of 166 cases of temporal arteritis reported in one series [1]. To the best of our knowledge, only one case of lateral medullary syndrome has been reported previously [4]. Our case serves to remind us of the differential diagnosis of stroke in the elderly, and the importance of considering this treatable cause, even in the absence of other constitutional symptoms or more classical presenting features.

Corneal ulceration has been previously reported in patients with giant cell arteritis in association with scleritis and is possibly related to ischaemia in the anterior segment of the eye [5]. In this case, there was no evidence of scleritis and the aetiology of the abrasion was more probably related to the corneal analgesia. Nonetheless, the ulcer was temporally associated with a rise in the ESR, and settled promptly with an increase in the steroid dose, raising ischaemia a possibility.

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