Short report

Giant cell arteritis with spinal cord infarction and basilar artery thrombosis

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Summary A patient with active giant cell arteritis developed paraparesis and dissociated sensory loss due to infarction in the anterior spinal artery territory at the level of T12. Three days later fatal basilar artery thrombosis occurred. No occlusive lesion was found to explain the anterior spinal artery syndrome but this was associated with active arteritis. Alternative possibilities are that thrombus was present in involved cervical feeding vessels, or that emboli arose from intimal involvement in larger vessels, or that the event was related to thrombocytosis.

Giant cell arteritis is an inflammation of medium and large arteries of unknown cause. The reported incidence ranges from 2.4⁻¹ to 9.3² cases per 100,000 per year depending upon the inclusion of biopsy positive cases only or additional biopsy negative cases; it is rare under the age of 55 years and there is a female preponderance of 2 to 1. It is often accompanied by headache, malaise, anorexia, weight loss, fever, symptoms of polymyalgia rheumatica and a risk of neurological complications due to occlusive thrombus in affected arteries. A panarteritis involves vessels focally or along their whole length. Initially there is an intimal proliferation of cellular tissue and mucoid intercellular substance containing a variety of inflammatory cells. There is fragmentation of the internal elastic lamina and granulomatous infiltration of the arterial wall, which is most marked in the media. Multinucleated giant cells lie in the intima or near breaks in necrotic media. There is evidence for immunity to elastic arterial tissue with an association between susceptibility and the amount of elastic tissue in the walls of involved arteries.³

The condition is best known for its involvement of cranial vessels particularly the external carotid, temporal and vertebral arteries but the aorta and its branches may be involved, the intracranial arteries to a lesser extent and spinal cord vessels least of all. Basilar artery thrombosis is a frequent cause of death. We believe this is the first report of a case with spinal cord infarction in the anterior spinal artery territory.

Case report

A 76-year-old man presented with six weeks severe generalised headache. He smoked twenty cigarettes per day and had chronic obstructive airways disease. On examination his blood pressure was 140/90 mm Hg, his temporal arteries were palpable but there was no scalp tenderness. The ESR was 84 mm hour, haemoglobin was 10-6 g/dl, PCV 0-33% mean corpuscular volume (MCV) 79 fl, mean corpuscular haemoglobin (MCH) 25-2 pg, blood white cell count (WBC) 9-1 × 10⁹/l, platelets 566 × 10⁹/l, urea 9-5 mmol/l, albumin 35 g/dl, total protein 81 g/dl, alkaline phosphatase 132 IV/l (30–100) and aspartate transaminase 41 IU/l (7–40).

He was given 80 mg oral prednisolone but one hour after the first dose he developed a sudden onset of low back pain and was unable to stand because of leg weakness. Muscle tone was reduced, there was symmetrical weakness of MRC grade 3 hip flexion, grade 4 hip extension and knee flexion, grade 4+ knee extension and ankle dorsiflexion and grade 5 plantar flexion. The leg reflexes were absent, the plantar responses were up-going, and there was a spinothalamic sensory loss to T₁₂, with preservation of touch, joint position and vibration sense. He developed urinary retention. A CT brain scan showed mild cerebral atrophy, later confirmed at necropsy, and water soluble contrast myelography was normal. Lumbar spinal fluid

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Fig (a) Temporal artery showing intimal proliferation, and irregularly damaged internal elastic lamina (arrows) (Elastic-van Giesen × 100) (b) Part of the vessel wall magnified to show multinucleate giant cells (small arrows) and the irregularly damaged elastica (large arrows). (× 250) (c) Transverse section of the anterior spinal cord at T12 level. Bilateral medial necrosis extends into the right anterior horn (arrow). (H and E × 63).
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contained 2 WBC/mm³, 20 RBC/mm³ and 0.39 g/l of protein. A diagnosis of anterior spinal artery territory cord infarction was made. High dose prednisolone was continued and the headache resolved after two days.

Three days later he suddenly felt unwell with vomiting, perspiration and drowsiness and over a period of three hours developed a right homonymous hemianopia, deviation of the eyes to the left and then to the right, left Horner’s syndrome, left lower motor neuron facial paralysis and decreasing consciousness followed by respiratory arrest.

At necropsy the right temporal artery was found to be pale, firm and tortuous. On microscopy there was severe temporal arteritis with intimal proliferation, fragmentation of the elastica and vessel wall inflammation with giant cells. (figs A and B) Major arteries including the aorta contained little atheroma and no other macroscopic changes. Examination of brain and spinal cord after fixation showed that vessels of the circle of Willis were free of atheroma and the lumen of the basilar artery contained thrombus in most of its length. Coronal brain sections showed slight ventricular enlargement, but there were no gross structural lesions. Microscopically there was perivascular rarefaction around vessels in frontal and parietal lobe white matter. In the brainstem there was infarction of the midbrain and pontine tegmentum with partial destruction of all tissue components with patches of extravasated blood. At this level the basilar artery showed damage and reduplication of the internal elastic lamina and a segment of adventitia with chronic inflammatory cell infiltration. Near its origin the lumen was filled with thrombus and the elastica was damaged. A paramedian branch also showed damaged elastica and inflammatory infiltration. The available sections did not show arteritis of vertebral arteries or other cranial vessels. Segments of cervical, thoracic and lumbar spinal cord were examined. At T12 there was partial infarction of the anterior columns involving the white matter on both sides and the medial half of the right anterior horn (fig C). The affected area showed infarction, small exudative haemorrhages and diffuse and focal inflammatory cell infiltrates. A vascular occlusive lesion could not be demonstrated at the levels examined.

Discussion

The patient had giant cell arteritis diagnosed clinically and confirmed at post mortem examination of the vessel. The events suggestive of brainstem infarction were due to thrombosis of the basilar artery. Drowsiness and coma is explained by ischaemia of the reticular activating system and the right hemianopia by involvement of the left posterior cerebral artery. Vomiting and left Horner’s syndrome are explained by ischaemia in cerebellar branches. Segmental effects at the level of the basilar thrombus were eye deviation attributed to involvement of a paramedian branch and left seventh nerve paralysis due to ischaemia of the anterior inferior cerebellar artery territory in the lower pons. The paraparesis and dissociated sensory loss were due to partial infarction in the anterior spinal artery territory, but it was not possible to determine whether the cord infarction resulted from thrombotic occlusion of the anterior spinal artery or of a radicular branch. Locating the site of occlusions in cases of spinal cord infarction can be difficult and infarction in this vascular watershed may be due to occlusive lesions at a distance, as in a cervical feeding vessel.

It is possible that cord infarction was due to incidental atheromatous disease but the relative freedom from atheroma and association with active arteritis are against this. Alternative considerations are fragment embolisation from the aorta or an event related to the platelet count of $599 \times 10^9/l$.

This high platelet count is consistent with the finding of high counts in large series associated with platelet production rates 1.7 times higher than observed in healthy subjects. Platelet counts of this magnitude are associated with transient ischaemic attacks and stroke although it is not entirely certain that thrombocytosis alone can cause thrombosis. However, it is important to consider that thrombocytosis could precipitate ischaemic episodes either by embolisation of platelet clumps from inflamed intimal surfaces or by the release of thromboxane A2 (TXA2) by activated platelets producing platelet aggregation and vasoconstriction. Enhanced platelet aggregation associated with increased TXA2 production is seen in the active phase of other arteritic conditions and could explain this previously unreported complication of giant cell arteritis and other complications, for example, ischaemic coronary events. In addition corticosteroids may increase transiently platelet numbers thereby creating a period of vulnerability to such events at the start of treatment. These points introduce the question of treatment either with cyclooxygenase inhibitors (aspirin) or thromboxane synthase inhibitors to accompany the first few days of steroid treatment of active giant cell arteritis.

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References


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