

Fatal vertebral giant cell arteritis

M M Sheehan, C Keohane, C Twomey

Abstract

Death due to giant cell arteritis (GCA) is rare, and is usually caused by coronary or vertebral arteritis in the acute phase of the disease. A case of fatal GCA is reported in a woman with a normal erythrocyte sedimentation rate, who had been treated for temporal arteritis for eight months. Post mortem examination showed a dissection and thrombosis of the intracranial portion of the left vertebral artery caused by giant cell arteritis. Focal coronary artery GCA was also found. As far as is known, this is the only case in which dissection of the vertebral artery attributable to GCA has been reported.

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Death from giant cell arteritis (GCA) is rare, and may be attributable to insufficient treatment.¹ Death is usually caused by coronary or vertebral arteritis occurring in the acute phase of the disease.^{1,2} Late mortality due to cerebral infarction is very rare.² Thrombosis associated with vasculitis represents the usual mechanism of infarction. While thrombosis was present in this case, considerable dissection of the vessel wall was also identified; this may have contributed to brain stem and cerebellar infarction.

Case report

An 85 year old woman presented with a five day history of occipital headache and bilateral tenderness over the temples. She had had poor vision for several years due to macular degeneration. Recent visual or other symptoms were absent. She was not receiving any medication and had been in good health. On examination she was afebrile, normotensive, and in sinus rhythm. She had palpable tender superficial temporal arteries. Neurological and general systemic examination yielded normal results. Her mental test score was 9 out of 10. On admission her ESR was 40 mm/first hour, but increased to 70 mm/hour one day later. Her haemoglobin concentration was 130 g/l, white cell count $9.24 \times 10^9/\mu\text{l}$ (neutrophils 75%, lymphocytes 13.7%) and the platelet count has $68 \times 10^9/\text{l}$. Chest x ray picture, electrocardiogram, electrolytes, and urinalysis were all normal.

Temporal arteritis was diagnosed and she was treated with prednisolone 60 mg/day.

She did not have a temporal artery biopsy. Substantial improvement occurred over the next two weeks and steroids were gradually reduced over the subsequent four months to a maintenance dose of 5 mg/day. She was reviewed monthly and remained asymptomatic, with an ESR ranging from 5-20 mm/hour (normal). Drug compliance was apparently good. Eight months after her initial presentation prednisolone treatment was reduced to 2 mg daily. Three weeks after this she was readmitted with a four day history of acute onset of confusion, vomiting, memory loss and inappropriate behaviour. On examination she was afebrile, normotensive, and in sinus rhythm, but was drowsy, poorly responsive to commands, and had a mental test score of 2 out of 10. She had reduced power in all four limbs, normal tone, and symmetrically brisk reflexes. The left plantar reflex was extensor. A left directional eye drift was noted. The ESR was 5-20 mm/hour. Haemoglobin concentration and white cell count were normal. It was not known if she had discontinued her treatment in the three weeks since her last review. A computed tomogram scan and arteriography were not performed. She became comatose and died two days later.

Pathology

Post mortem examination showed recent lateral medullary infarction and haemorrhagic infarction of the superior surface of the left cerebellar hemisphere due to thrombosis of the intracranial portion of the left vertebral artery (fig 1). The thrombus was focal at the site of the inflamed dissected wall and extended for about 1.5 cm. It did not extend throughout the full length of the vertebral artery or into the basilar artery. The entire circle of Willis was examined histologically and sections were stained with haematoxylin and eosin, haematoxylin van Gieson, Verhoeff van Gieson, and Congo red. Multiple levels were cut. Histological examination of the left vertebral artery showed disruption of the internal elastic lamina surrounded by multinucleated giant cells and inflammatory cells throughout the tunica media. There was a dissection at the level of the fragmented internal elastic lamina at a site where multiple giant cells and inflammatory cells were present (fig 2). There were only scanty erythrocytes in the dissected media but hyaline acellular material was present, similar to that in the degenerated elastic lamina. The

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Figure 1 Vertebral artery showing thickened inflamed wall, with dissection (D) at the level of the degenerated internal elastic lamina, surrounded by inflammatory cells. A multinucleated giant cell is seen at the edge of the dissection (arrow). Retracted thrombus is present within the vessel lumen (haematoxylin and eosin). L = lumen; I = Intima; M = Media.

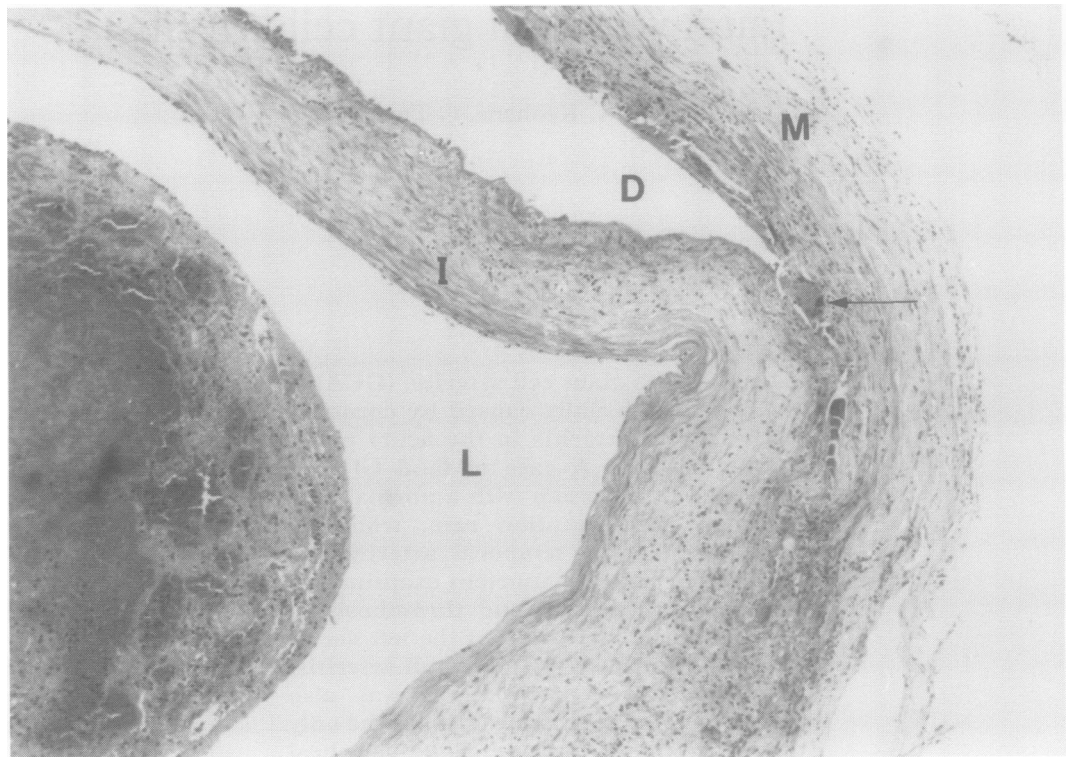
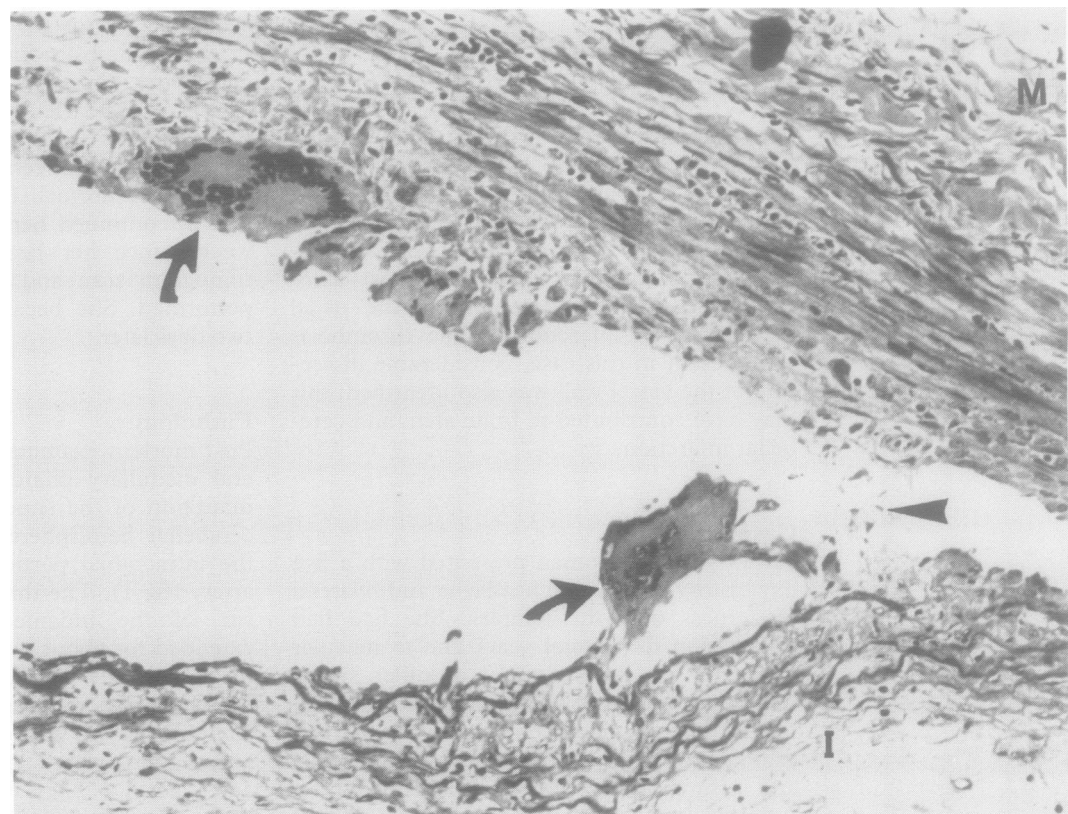


Figure 2 High power of dissection showing the multinucleated giant cells (arrows) within and lining the dissection and inflammatory cells scattered through the wall. Scanty erythrocytes and leucocytes are present in the dissection (arrowhead). The internal elastic lamina has fragmented (Verhoeff van Gieson). I = Intima; M = Media.



dissection extended over a distance of about 2.5 cm and was present proximal and distal to the thrombus. There were no granulomas or amyloid. The intracranial portion of the right vertebral artery, the basilar artery, and remaining branches of the circle of Willis and meningeal vessels were not inflamed. There was an old cavitated left parietal infarct with telangiectases, hyalinised arterioles, and gliosis.

The heart was normal on macroscopic

examination, but histological examination showed a small area of myocardial necrosis, less than 48 hours old, in a random section of the wall of the right ventricle. The right main coronary artery showed focal GCA but without thrombosis or occlusion. The left coronary, renal, thyroid, pulmonary and pancreatico-splenic arteries were not inflamed. The temporal and ophthalmic arteries were not examined. No opportunistic infections were present.

Discussion

Giant cell arteritis is a relatively common disease in the elderly¹ and can affect almost all the large arteries, including the aorta.^{3,4} The superficial temporal and ophthalmic arteries are often affected, causing headache and visual disturbance. Polymyalgia rheumatica frequently accompanies these symptoms. The intracranial arteries are rarely affected,^{3,4} but this has been reported in some cases.^{1,5}

The pathogenesis is unknown.^{3,4} Fragmentation of the internal elastic lamina, an inflammatory infiltrate of mixed mononuclear and polymorphonuclear leucocytes, and characteristic multinucleated giant cells are the histological hallmarks of the disease.⁴ Immunological mechanisms have been postulated but the precise pathogenic processes have not been elucidated.³ Survival rates for GCA are similar to those of an age matched population,¹ and fatal complications in well treated patients are rare, even in those with chronic relapsing disease.² Early mortality (within six weeks of presentation)¹ while uncommon, is usually associated with arteritis and brain stem infarction, or coronary arteritis and myocardial infarction.^{1,2} Late mortality (more than six weeks after presentation) due to cerebral infarction is very rare.² Death has been attributed to insufficient corticosteroid treatment.¹ In fatal cases due to vertebro-

basilar disease, necropsy showed either intracranial and extracranial arteritis with thrombosis¹ or extracranial vertebral arteritis, with unaffected intracranial vessels.¹

Our case is the only one in which dissection of the intracranial portion of the vertebral artery due to GCA is recorded, although dissection of the aorta has been documented before.¹ It is not clear whether our patient had continuous active arteritis or a relapse following reduction of her maintenance treatment. It is clear, however, that severe GCA with fatal complications occurred with a normal ESR, without recurrence of initial symptoms, and in spite of eight months of continuous treatment. This case also shows that active GCA may cause microscopic vertebral artery dissection.

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Renal failure caused by leukaemic infiltration in chronic lymphocytic leukaemia

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Abstract

A case of B-CLL which was complicated by chronic renal failure due to leukaemic infiltration of the kidney is reported. Treatment with chlorambucil, prednisolone, and renal bed irradiation resulted in a substantial improvement in renal function which persisted until the the patient's death from marrow failure some eight years later. The temporal association between treatments and response suggested that renal bed radiotherapy had contributed to the improvement in renal function. This case is one of only two reported cases in which chronic renal failure due to CLL has been treated with radiotherapy. It is unique in that the renal response was shown histologically.

Leukaemic infiltration of the kidney is common in CLL but, characteristically, is not associated with renal impairment. An improvement in renal function has

been described in two patients with acute renal failure after chemotherapy.

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Case report

A 60 year old man presented with generalised lymphadenopathy without other abnormal physical signs. Investigation showed the following: haemoglobin 152 g/l; white cell count $11.3 \times 10^9/l$ (lymphocytes $6.8 \times 10^9/l$); and platelet count $120 \times 10^9/l$. The peripheral blood film showed a lymphocytosis with occasional "smudge" cells. Peripheral blood lymphocyte surface markers were consistent with B cell CLL (91% positive for HLA-DR, 81% positive for CD19, 91% positive for CD5, 9% positive for CD2, 5% weakly positive for SmIg, 0.5% positive for FMC7, 1% positive for CD10, 3% positive for CD25). A bone marrow aspirate showed that 65% of nucleated cells were small lymphocytes. A bone

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