Discussion

Giant cell arteritis is a relatively common disease in the elderly and can affect almost all the large arteries, including the aorta. 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, but this has been reported in some cases.

The pathogenesis is unknown. 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. Immunological mechanisms have been postulated but the precise pathogenic processes have not been elucidated. Survival rates for GCA are similar to those of an age matched population, and fatal complications in well-treated patients are rare, even in those with chronic relapsing disease. Early mortality (within six weeks of presentation) while uncommon, is usually associated with arteritis and brain stem infarction, or coronary arteritis and myocardial infarction. Late mortality (more than six weeks after presentation) due to cerebral infarction is very rare. Death has been attributed to insufficient corticosteroid treatment. In fatal cases due to vertebral-basilar disease, necropsy showed either intracranial and extracranial arteritis with thrombosis or extracranial vertebral arteritis, with unaffected intracranial vessels.

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. 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.


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 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 GCA 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 × 10⁹/l (lymphocytes 6.8 × 10⁹/l); and platelet count 120 × 10⁹/l. The peripheral blood film showed a lymphocytosis with occasional "smudge" cells. Peripheral blood lymphocytes 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
the preceding three weeks. Examination showed generalised lymphadenopathy without splenomegaly, mild hypertension (blood pressure 150/100 mm Hg) and grade II hypertensive retinopathy. Investigation showed: haemoglobin 106 g/l; white cell count 90.5 × 10⁹/l (lymphocytes 81.5 × 10⁹/l); a platelet count of 153 × 10⁹/l; urea of 27.8 mmol/l; creatinine of 563 μmol; creatinine clearance 15 ml/minute; and a urinary protein excretion of 0.7 g/day. Serum calcium and uric acid values remained normal. Moderate glycosuria was noted on urinalysis in the presence of a plasma glucose concentration of 4.8 mmol/l.

A renal ultrasound scan showed normal sized kidneys with no evidence of obstructive nephropathy; the only abnormality on plain tomography was the presence of slightly irregular renal outlines. A needle renal biopsy specimen showed severe tubular atrophy with wide separation of tubules by interstitial connective tissue heavily infiltrated by small lymphocytes exhibiting the same surface marker pattern as the peripheral blood lymphocytes. The infiltrate was more pronounced in the cortex than in the medulla. Twenty nine of 33 glomeruli showed ischaemic collapse with periglomerular fibrosis, the remainder showing global sclerosis. The arteries showed moderate fibroelastic intimal proliferation and mild hyaline arteriolar sclerosis was present (fig 1). These appearances were interpreted as showing severe ischaemic nephrosclerosis with infiltration of the cortical interstitium by CLL.

The patient received oral chlorambucil 20 mg daily for two days with oral Allopurinol 200 mg daily. Treatment for renal failure consisted of a low protein diet and oral bicarbonate. Atenolol 50 mg daily was given for the hypertension. Three weeks later the lymphocyte count had fallen to 61 × 10⁹/l but there had been a substantial decline in renal function with urea reaching 31.9 mmol/l and creatinine 1124 μmol/l. Further oral treatment with chlorambucil 20 mg once a day for three days and prednisolone 20 mg three times a day for 10 days was given in addition to low dose renal bed irradiation (400 cGy in two fractions given on successive days). Over the next two weeks the serum creatinine concentration fell to 496 μmol/l and the urea to 17.9 mmol/l. This was followed by a fall in the lymphocyte count to 14.5 × 10⁹/l at about five weeks after the second course of chemotherapy. Figure 2 shows the changes in serum creatinine and lymphocyte count in relation to treatment during this period.

Treatment for CLL was continued with monthly courses of chlorambucil for a total of nine courses. One month after completing chemotherapy the patient was asymptomatic, with lymphadenopathy confined to two small cervical nodes. The lymphocyte count was normal at 2.6 × 10⁹/l and renal function had shown further slight improvement with a serum creatinine of 362 μmol/l and urea of 16.1 mmol/l. Renal biopsy was repeated five months after radiotherapy and this showed a pronounced reduction in the extent of...
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Renal biopsy specimen taken after radiotherapy showing a less extensive infiltrate. Tubular atrophy is still apparent and there is periglomerular fibrosis.