Sir,

Positive temporal artery biopsy in a patient on therapeutic doses of steroids for six years

Temporal arteritis can be a challenging disease both to treat and to diagnose. The diagnosis is made on clinical suspicion supported by a raised ESR and can be confirmed with a positive biopsy result.[1] We report a case in which the patient had been treated for 6 years on steroids, and who underwent a temporal artery biopsy which illustrated characteristic features of active arteritis.

Case report

This 69-year-old woman presented in 1993 with a history of nausea, general malaise, sore neck, anorexia and weight loss. There were no visual symptoms and no jaw claudication. ESR was >120 mm/h and she was not anaemic. C3 was raised at 191 mg/dl; C4 was within normal limits. U&E, renal function, liver function, plasma protein and protein electrophoresis were all normal. Rheumatoid factor, ANA and ANCA were negative. She was examined by a consultant physician and, in view of the constitutional symptoms and high ESR, a diagnosis of polymyalgia rheumatica was made and she was commenced on 20 mg of oral prednisolone daily.

Over the next 3 months the dose of steroids was gradually reduced to 12.5 mg. She complained of a more severe temporal headache and an episode of visual disturbance. On examination she was found to have temporal artery tenderness and an ESR of 88 mm/h. A diagnosis of temporal arteritis was made, the dose of oral steroids was increased to 60 mg and her symptoms improved. The option of a temporal artery biopsy was discussed with the patient but she declined this intervention.

Over the next 5 years she was reviewed by a number of consultant physicians, all of whom on balance felt that the patient was suffering from temporal arteritis. The patient suffered significant side-effects from the steroids including gastrointestinal upset, weight gain and memory loss. Reduction of steroids resulted in re-occurrence of her temporal arteritis symptoms. Steroid-sparing medications in the form of azathioprine, methotrexate and cyclosporin were attempted but all withdrawn due to intolerance.

The patient's condition continued to be a challenging management problem. In September 1999, just over 6 years since the initial presentation and the institution of continuous steroid treatment, she was referred to the ophthalmology department with a request for temporal artery biopsy. At this time the ESR was 12 mm/h.

The biopsy specimen was a segment of temporal artery 8 mm long by 1 mm in diameter. The specimen was divided and embedded in paraffin wax. H&E microscopy confirmed the presence of a medium-sized artery. Fibrointimal thickening with a chronic inflammatory cell infiltrate was present which was most marked towards the adventitia. A well-formed epithelioid granuloma was present within the adventitia, and the appearances were reported as being consistent with giant cell (temporal) arteritis (Fig. 1).

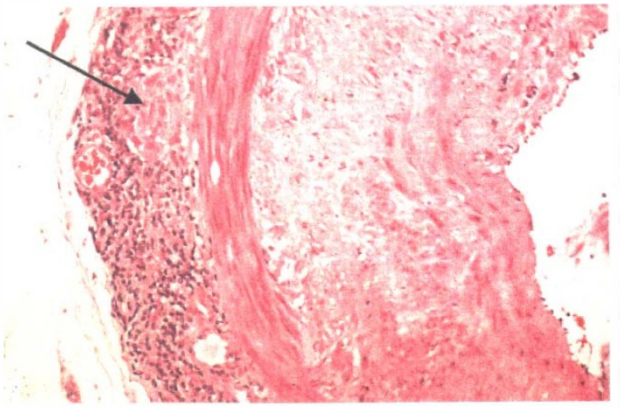

Fig. 1. *Temporal artery specimen stained with H&E (×160). Fibrointimal thickening with chronic inflammatory cell infiltrate was present which was most marked towards the adventitia. Arrow indicates epithelioid granuloma present in the adventitia. The appearances were reported as being consistent with giant cell (temporal) arteritis.*

Comment

A positive temporal artery biopsy result confirms the diagnosis of temporal arteritis and justifies the use of long-term steroid or other immunosuppressive treatment.[2] A negative biopsy does not exclude the diagnosis of temporal arteritis due to the focal and segmental distribution of the inflammation in an affected artery. Discussion has surrounded the desired length of the biopsy[3] and the efficacy of a bilateral biopsy.[4] The histological features of temporal arteritis can be differentiated from other forms of vasculitis. In active disease transmural inflammation with prominent macrophages and giant cells, disrupted internal elastic lamina and more rarely necrotising change and thrombus formation are seen.[5,6] Healed arteritis is characterised by marked fibrosis, and disruption of the internal elastic lamina with no inflammatory cells.

Traditional teaching has encouraged the prompt acquisition of a biopsy with the rationale that sufficient immunosuppression will rapidly modify the inflammatory findings (mirroring the improvements in clinical symptoms). What is the evidence supporting this action? The current literature reviewed gives conflicting results on the effect of the steroid treatment on the rate of positive biopsy results. One study[7] reviewed 535 consecutive patients and recorded the dose and duration of steroid treatment received before temporal artery biopsy. The authors concluded that they could not prove that histological features were affected by prior steroid treatment and that positive biopsies were found even after more than 14 days of treatment. The findings of a second group[8] conflict with this conclusion. This study reviewed 132 cases, and concluded that the yield of positive biopsy samples fell significantly after only 1

week of treatment. Several anecdotal reports have illustrated positive results after longer periods of therapeutic levels of steroids: 1 month[9] and 6 months.[10]

Does the finding of positive active temporal arteritis despite steroids suggest insufficient immunosuppression, or that the histological findings do not necessarily correlate with clinical activity? This latter suggestion was made in a paper which took a second biopsy in 20 patients after completion of treatment for temporal arteritis.[11] The group reported that although there was some correlation between the clinical and histological signs of active disease during and after treatment, this was not complete.

The presence of a positive biopsy in this case has confirmed the diagnosis of temporal arteritis. This case at least anecdotally suggests that irrespective of the duration of the steroids temporal artery biopsy may be helpful and should be considered. It is difficult to be confident of the significance of this result in terms of the patient's disease stage. Ultimately, her systemic treatment will be titrated against her symptoms. A positive temporal artery biopsy result when the patient first presented with symptoms suggestive of an arteritis would have confirmed the need for immunosuppression from the outset. This histological confirmation becomes particularly important in patients who develop complications associated with immunosuppression.

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Sir,

Sphenoid dysplasia and temporal lobe prolapse in neurofibromatosis type 1

The neurofibromatoses are autosomal dominant disorders of the nervous system that primarily affect the development and growth of neural cell tissues resulting in neural, skeletal and dermatological abnormalities. Ocular involvement is manifold and may involve the eyelids, cornea, iris, retina, choroid, optic nerve and the bony orbit. We present a case of an asymptomatic patient with plexiform neuroma in whom ipsilateral sphenoid dysplasia led to herniation of intracranial structures into the orbit.

Case report

A 14-month-old female infant was referred to the eye clinic by a health visitor with a swelling over the left side of the face involving the eyelid. The mother had noticed it at birth and attributed it to the forceps delivery, which had also caused significant bruising of both cheeks. Post-natal history and milestones were normal. There was no significant family history.

Ocular evaulation revealed a visual acuity of 6/7.5 using the Cardiff cards with both eyes open. A non-tender soft tissue swelling with brownish discoloration was noted in the left peri-orbital region. The swelling extended from the lateral aspect of the upper eyelid onto the zygoma (Fig. 1). Anterior segment was normal and corneal reflexes were central. Pupillary responses were

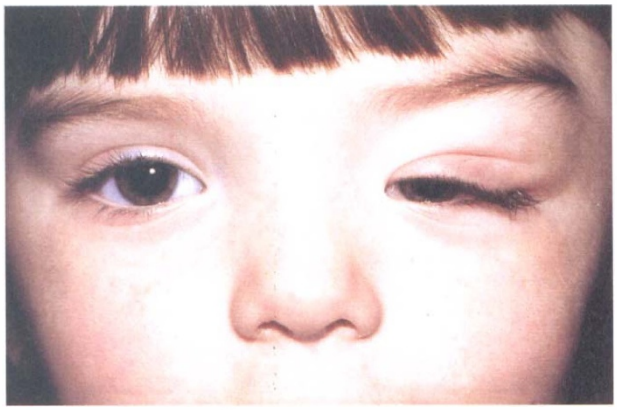

Fig. 1. *Photograph showing the swelling in the left upper lid and the resultant contour of the lid.*