Case report

Development of giant cell (temporal) arteritis in a patient 'adequately' treated for polymyalgia rheumatica

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SUMMARY A 70-year-old woman presenting with typical polymyalgia rheumatica (PMR) and a normal temporal artery biopsy appeared to respond completely to low-dose prednisone therapy. A subsequent biopsy showing temporal arteritis with a normal sedimentation rate and no recurrence of myalgic symptoms emphasizes the unpredictable course of treated PMR and the need for continued clinical as well as laboratory follow-up.

Polymyalgia rheumatica (PMR) (Kogstad, 1965; Bruk, 1967; Hamilton *et al.*, 1971) is typically associated with a raised erythrocyte sedimentation rate (ESR) and frequently responds to corticosteroid treatment in low dosage. Giant cell arteritis (GCA) may complicate PMR in up to 50% of patients (Hunder *et al.*, 1969; Fauchald *et al.*, 1972; Hamrin, 1972). PMR and GCA rarely occur with a normal ESR; and the occasional patient who has developed or has had an exacerbation of GCA while undergoing treatment with corticosteroids has also had a concomitant rise in ESR (Kogstad, 1965; Beevers *et al.*, 1973; Klein *et al.*, 1975). We report a patient with typical PMR who developed temporal arteritis despite apparently adequate corticosteroid therapy and a normal ESR.

Case report

A 70-year-old White female presented with a 3-month history of morning stiffness and pains of the hips, shoulders, fingers, and wrists, and intermittent fever to 37.8°C. Symptoms improved during a 1-week trial of oral corticosteroids but recurred after the medicine was discontinued.

The temporal arteries were normal. A grade II/VI systolic murmur was heard at the left lower sternal

border. Musculoskeletal examination showed the proximal arm muscles were tender and range of motion of the shoulders, wrists, hips, and knees was decreased without evidence of synovitis. Consecutive Westergren ESRs were 76, 90, and 83 mm/h, haemoglobin 12.6 g/dl, haematocrit 38.4%, and WBC 7800/mm³ ($7.8 \times 10^9/l$). Serum alkaline phosphatase was 121 IU/ml (normal 4-85) and lactic dehydrogenase 211 IU/ml (normal 90-200), with normal SGOT and bilirubin. Tests for antinuclear antibody, LE cells, and rheumatoid factor were negative. Complement β_1c (Hyland Plate) was 3.7 g/l (normal 0.97-1.89). X-rays of hands, wrists, knees, and shoulders showed only minimal osteoporosis. An electrocardiogram was normal, as was a left temporal artery biopsy (Fig. a). Right deltoid muscle biopsy showed changes of atrophy, and variation in size and splitting, mainly in type II fibres.

She was treated with prednisone 5 mg twice daily with subsidence of symptoms in one week, and return to normal of the ESR associated with a rise in haemoglobin to 13.6 g/dl and haematocrit to 41% over 2 months. Prednisone dosage was gradually reduced over the next year to 5 mg in the morning. ESR was 15 mm/h, and dosage was decreased to 5 mg and 4 mg on alternate days. At this time she developed angina of effort.

Two months later she was admitted to hospital with a 2-week history of sharp pains in the temples and a tender enlargement of the right temporal

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Fig. Temporal artery biopsies. (a) Left temporal artery biopsied in October 1973 (Haematoxylin-eosin $\times 45$) is normal.

(b) Right temporal artery biopsied in March 1975 (Haematoxylin-eosin $\times 45$) shows marked inflammation with obliteration of the vessel lumen, round cell infiltration, destruction of normal architecture of vessel wall, and occasional giant cells.

artery. Westergren ESR was 20, 19, and 20 mm/h, and haemoglobin 13.4 g/dl. A right temporal artery biopsy (Fig. b) showed typical features of giant cell arteritis and a left deltoid muscle biopsy was similar to the previous one. Prednisone 10 mg twice daily rapidly alleviated her temporal pain. Subsequently, she has maintained a normal ESR and remained asymptomatic except for continued angina pectoris.

Discussion

The patient's initial presentation was entirely consistent with uncomplicated PMR. Clinical symptoms were typical, the ESR was markedly raised, and she responded to low-dose corticosteroids with relief of symptoms and by a return of the ESR to normal. At that time there was no evidence of giant cell arteritis either clinically or by temporal artery biopsy. The development of angina pectoris was thought to represent atherosclerotic cardiac vascular disease, although involvement of the coronary arteries by the arteritis has been reported by others (Morrison and Abitbol, 1955).

Development of giant cell arteritis occurred as the prednisone dosage was slowly being reduced but with a normal ESR and no recurrence of myalgic symptoms. Others have noted the hazards of relapse in the course of GCA when corticosteroid therapy has been discontinued or reduced too rapidly (Hamilton *et al.*, 1971; Klein *et al.*, 1975). Relapses have also occurred with very gradual reduction of corticosteroid dosage or during long-term maintenance therapy, although the ESR was raised at these times (Beevers, *et al.*, 1973; Klein *et al.*, 1975). The ESR is not always raised in patients presenting with PMR (Bruk, 1967). However, it does seem unusual for a patient on steroid therapy to have a relapse of PMR or manifest symptoms of GCA without increase in the ESR. Beevers suggested that this could occur but offered no evidence, though he did note 5 symptomatic recurrences with only modestly raised ESRs of <45 mm/h (Beevers *et al.*, 1973).

The failure of our patient's ESR to rise above

20 mm/h on repeated determinations at the time of her presentation with clinical and biopsy-proven temporal arteritis appears to be distinctly uncommon, and emphasizes again the need for prolonged intensive follow-up of symptoms and ESR, as well as the potential development of GCA in patients with apparent adequate treatment for PMR.

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