Case report

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

RICHARD I. RYNES, PHILIP MIKA, AND LEE E. BARTHOLOMEW
From the Department of Medicine, Albany Medical College, and Albany Medical Center Hospital, Albany, New York

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 x 10⁹/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 β₁c (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 IIB 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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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 Abibol, 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.

We thank Dr. Peggy Hanson, Department of Neurology, Albany Medical College, for processing and interpreting the muscle biopsies.

References


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doi: 10.1136/ard.36.1.88

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