Polymyalgia Rheumatica

A. St J. DIXON, M D, F R C P, Physician, St Martin's Hospital, and Royal National Hospital for Rheumatic Diseases, Bath

Polymyalgia rheumatica was first described under the name of 'senile rheumatic gout' by Bruce in 1888, then as a 'myalgic syndrome of the elderly' by Kersley in 1951 and 'anarthritic rheumatoid disease' by Bagratuni in 1953. In France, Forestier and Certonciny (1953) called it 'pseudo-spondylite rhizomelique', but it is Barber's (1954) name, polymyalgia rheumatica, that has become accepted.

Polymyalgia rheumatica can be defined as 'a protracted disease state of the elderly characterised by proximal pain and stiffness, general illness, mild anaemia, and a raised sedimentation rate'. The diagnosis is made mainly by exclusion, as indicated by the way in which my own series of cases has been accumulated.

Since 1962 a Westergren sedimentation rate has been measured in all new referrals to my out-patients (Dixon *et al.*, 1966). Rates over 30 mm/hr were repeated for confirmation and patients with rates repeatedly over 40 mm/hr were extensively investigated to eliminate serious occult disease such as carcinomatosis, myelomatosis, leukaemia, and 'collagen diseases'. There

TABLE 1. Polymyalgia Rheumatica and Temporal Arteritis

	West London	Bath	Total
Male	5	6	11
Female	26	28	54
	31	34	65

remained a group of elderly patients who had general illness, morning stiffness, aches and pains, and a persistently raised sedimentation rate, with no obvious cause. A therapeutic test using ACTH or prednisolone was carried out on these patients, and those who responded rapidly were diagnosed as having 'polymyalgia rheumatica'.

The point has to be made that polymyalgia rheumatica and the myalgic prodromal phase of temporal arteritis are clinically indistinguishable, and the two conditions should therefore be considered together. Both are diseases of the elderly and both are increasingly common.

This series of sixty-five cases is drawn from the West London area and from Bath (Table 1). Both are retirement areas with a higher than national average proportion of women over sixty years old. All of the West London series had routine temporal artery biopsies done—whether or not there was evidence of temporal arteritis. At Bath, biopsy has only been done in cases of clinical doubt. There was one new patient for every ten new referrals for rheumatoid arthritis and for every single new referral for gout. Most of the patients were about sixty years of age, the eldest being ninety-two. What one observes, of course, depends on one's viewpoint. Because my viewpoint is that of a general physician with a bias to rheumatology, my patients get referred for rheumatic pains and difficulty in moving their limbs. Specialists in other fields might have a different experience to report.

CLINICAL PICTURE

Polymyalgia rheumatica has no specific sign or pathological test, but nevertheless the general clinical picture is often so characteristic that the diagnosis can be made on the telephone. The patient, usually a woman over sixty, notices the onset, often quite rapid, of 'rheumatic' symptoms. She feels generally unwell, stiffens up easily, finds movement painful. Stiffness, pains, and tenderness are proximal, being felt in the muscles around the shoulders, neck, spine, hips, and thighs, in contrast to the stiffness and pain of rheumatoid arthritis. Initially they may be asymmetrical but later affect both sides equally. The pain may become 'agonising', the patient fearing even physical examination as this involves movement of the limbs.

Perhaps the most striking symptom is morning stiffness, similar to that of rheumatoid arthritis but much more severe and without joint swelling. One patient was quite unable to get out of bed in the morning; by 11 a.m. she could manage fairly normally. A sixty-six year old retired matron was almost paralysed in the morning by stiffness and pain when she first suffered from polymyalgia rheumatica. Her sister would bring her a cup of tea in the morning, but she could not sit up to drink it, so it was put on the floor by the bed. Very gradually she would ease herself out of bed, and slide down on to the carpet. By resting face down on her elbows she could manage to get the cup to her lips. Two patients have attempted by sleeping all night in a chair to avoid this morning stiffness.

A true inflammatory arthritis affecting the shoulders, knees and occasionally the wrists has been seen long after the onset of the disease in four of the sixty-five cases. It is transient, not severe, not associated with radiological evidence of bone erosions and recovers completely. It does not lead to deformity. Some degree of osteoarthrosis is inevitable in this age group, but is

not related to general illness and associated depression. Thus, the two most common misdiagnoses are osteoarthrosis and neurosis. The only permanent joint change is a mild sclerosis of the sacroiliac joints, which Andrews (1965) has reported. There has been only one example in this series.

RELATIONSHIP TO TEMPORAL ARTERITIS

Four patients with this sequence of the clinical picture of polymyalgia rheumatica and no signs of arteritis, despite very careful testing (including biopsy), have developed temporal arteritis at a later date. These are in addition to those patients who gave a history of myalgic symptoms before they presented with temporal arteritis. A typical history is as follows. Mrs E. K., aged seventy, developed pains in the right groin in 1963, spreading to the shoulders and both thighs. In 1964 she was admitted to hospital and the temporal arteries were recorded as normal. She recovered with ACTH. Four months later, still taking ACTH, but not in a dose sufficient to suppress the raised sedimentation rate, she developed headaches, could not focus and her jaws became tender. At this stage the temporal artery pulsation disappeared and the biopsy of the artery was positive. In these and similar instances temporal arteritis has been observed to develop in patients presenting with polymyalgia rheumatica, usually when the standard dose is reduced or is inadequate to suppress the sedimentation rate.

TEMPORAL ARTERITIS

Other names for this condition are cranial arteritis, senile arteritis, giant cell arteritis, and Horton's arteritis (Horton *et al.*, 1934). The term 'polymyalgia arteritica' (Hamrin *et al.*, 1964) emphasises the cases with both myalgic symptoms and arterial involvement. The classical description emphasises the tender thickening and beading of the temporal artery. Pulsation, it is said, disappears later. My experience has been that while such cases do occur, it is much more common for patients with polymyalgic symptoms and a high sedimentation rate either to have a clinically normal temporal artery, or else to show a primary loss of pulsation without going through any phase of tender thickening. This was verified in the West London series by routine temporal artery biopsies. About a third of these were positive, sometimes when there were no symptoms or signs pointing to involvement of the cranial arteries. Therefore, any reported series of polymyalgia rheumatica will certainly underestimate the frequency of arteritis if routine biopsies are not done. Moreover, it will underestimate the frequency if there is inadequate follow-up. It may take three or more years between the onset of polymyalgic disease and the development of the clinical picture of temporal artery

inflammation. Like other authors, we have done a number of muscle biopsies, electromyograms, serum creatinine kinase estimations, etc., with negative results. The only revealing investigation is the temporal artery biopsy. When pulsation has disappeared the artery may be difficult to find on biopsy, but this, in itself, is almost diagnostic. The pathologist should always be asked to stain for elastic tissue—the remnants of the artery may be discernible with this stain when the routine stain does not show it. Allowing the biopsy specimen to dry slightly on to a piece of card before dropping the card into the formalin to fix it stops the artery from curling up. This helps the histologist to orientate the specimen and get a good transverse section.

Histology

The typical appearance of the biopsy is of gross thickening of the walls of the vessels with loss of the normal distinction between intima, media, and adventitia and the appearance of giant cells in the walls. However, giant cells are not essential to the diagnosis and any granulomatous arteritis with or without thrombosis of the lumen is consistent with the diagnosis.

Prediction of a Positive Biopsy

Certain symptoms and signs have been found helpful in predicting a positive temporal artery biopsy. Scalp pain and tenderness may present in a number of ways, the patient being unable to brush the hair or to tolerate the pressure of spectacles. It is often unilateral and quite different from an ordinary headache. Visual disturbances, flashes of light, transient diplopia, and eye pain may indicate ophthalmic artery involvement. Claudication of the jaw, i.e. pain in the masseter muscle on chewing, is very characteristic. One patient could not get through a meal of roast duck, although he could manage Scotch broth easily. Absence of pulsation in the whole or part of the artery was more common than tender thickening or beading. A sensitive carotid sinus was quite often found: only light pressure on the carotid sinus will cause asystole of two or more beats. The patient must be lying flat before testing this, because of the possibility of fainting.

The Danger of Blindness

Inspection of the retina in two patients revealed partial retinal artery block. Such patients may develop complete retinal artery block with permanent blindness if the diagnosis is missed and the condition not treated. There should be no excuse for the tragedy exemplified by the following patient—a woman, aged sixty-nine, was seen by an experienced specialist in rheumatic diseases.

He found nothing much wrong to explain her complaint of pains in the neck, shoulders and back and sent her for physiotherapy. No sedimentation rate was done. However, she did not improve, and her doctor referred her to an experienced general physician who investigated her without being able to make a diagnosis. Again, no sedimentation rate was ordered. The patient was sent to a convalescent home where she developed sudden bilateral irreversible blindness. On readmission to hospital she was found to have a positive temporal artery biopsy and a sedimentation rate of over 100 mm. It is difficult not to believe that an earlier measurement of sedimentation rate would not have alerted suspicion and saved the patient's sight.

Temporal arteriography (Gillanders *et al.*, 1969) has confirmed that involvement of the cranial arteries is segmental. Thus, a patient may have arteritis yet show a negative temporal artery biopsy. Nevertheless, the temporal artery is the artery most frequently affected and most likely to show a positive biopsy, but clearly a negative biopsy cannot be relied upon entirely to rule out the danger of blindness.

None of my cases has developed blindness on treatment, even those known to have retinal artery involvement.

Regular Supervision of Treatment

The key investigation is the sedimentation rate. Rates of over 50 mm are the rule and over 100 mm are not uncommon. There is a tendency for those patients who subsequently show temporal arteritis to have rather higher rates initially. The aim of management is to give prednisolone in doses sufficient to suppress the sedimentation rate to a safe level. Arbitrarily, this is below 20 mm in one hour for men and below 30 mm in one hour for women. Patients are brought back routinely to the out-patient department so as to 'titrate' the dose against the sedimentation rate. In this way, we keep the dose to a minimum consistent with safety. Although the patient usually knows when the dose is inadequate (by return of stiffness), it is essential to realise that an inadequate dose may keep them feeling perfectly comfortable, yet be insufficient to suppress the sedimentation rate and thus suppress the development of temporal arteritis.

Related Syndromes

Some other syndromes in elderly people are probably related. These include a shoulder/hand syndrome with a high sedimentation rate in elderly men; a prolonged illness and unexplained normochromic anaemia with a haemoglobin of 50 per cent or less with a high sedimentation rate, responding

rapidly to prednisolone; and an acute sero-negative polyarthritis in elderly men with an excellent prognosis despite widespread initial joint involvement. These are additional to the bizarre presentations of giant cell arteritis such as scalp necrosis, tongue ulcer, cerebrovascular accidents associated with high sedimentation rate, and elderly versions of Pulseless Disease, resembling the Takayushu syndrome of young women. Doubtless there are others.

DIFFERENTIAL DIAGNOSIS

Lack of an independent single essential sign, test, or aetiological agent for this group of conditions makes it all the more necessary to exclude other diseases that have a different treatment or prognosis. The list of these could be very long—but the most likely conditions to present in a similar way can be divided into two groups, those with and those without a raised sedimentation rate. Those with a raised sedimentation rate include the prodromal phases of rheumatoid arthritis and disseminated lupus erythematosus, multiple myelomatosis, carcinomatosis with multiple secondary deposits or neuromyopathy, polymyositis, polychondritis and thyrotoxic myopathy associated with autoimmune thyroid disease. In younger patients the prodromal phase of erythema nodosum and ankylosing spondylitis may be similar.

Those with a normal sedimentation rate include myxoedema, 'stiff-man syndrome', chronic brucellosis and, occasionally, tabes dorsalis, myotonia, and Parkinsonism. Four of my patients (not included in the series) were initially thought to have polymyalgia rheumatica but had not. Two subsequently developed rheumatoid arthritis and two had occult infections, one of which was tuberculous.

COMPLICATIONS OF TREATMENT

The management of polymyalgia rheumatica is essentially prednisolone therapy under regular supervision. Corticotrophin has been unsatisfactory, as it has caused dangerous fluid retention in some patients. There is no set dose of prednisolone. The dose is the dose that works and it has to be given as long as the sedimentation rate is raised without it. This may mean treatment for many years. I have the strong impression that not only do these patients respond dramatically and consistently to prednisolone, but they do not seem to develop the usual adverse effects. (The only complication of any frequency has been diabetes.) It is as though one were dealing with a deficiency state. A comparable group of elderly patients with rheumatoid

arthritis, given corticosteroids for several years, would certainly be troubled by osteoporosis, peptic ulcers, and purpura.

PROGNOSIS

The prognosis is very variable from eighteen months to over fourteen years. The disease does not appear to shorten life, although some of the patients in the series have now died of unrelated causes.

CONCLUSION

Two 'medical mottoes', neither of them original, have been helpful:

1. 'If you suspect temporal arteritis give prednisolone and then arrange for temporal artery biopsy.' You may save the patient's eyesight, as blindness can come on with extraordinary rapidity if the retinal arteries are affected.
2. 'No elderly patient with a raised sedimentation rate should be allowed to die undiagnosed without the exhibition of steroids.'

This article is based on a paper read at the Rheumatology Conference held at the Royal College of Physicians in May 1969.

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