Fortnightly review

Polymyalgia rheumatica and temporal arteritis: diagnosis and management

A J Swannell

Polymyalgia rheumatica and temporal arteritis are regarded as clinical syndromes affecting elderly people. They may occur in the same patient, producing constitutional symptoms with increased acute phase reactants. Both syndromes respond rapidly to corticosteroids, and in both syndromes temporal artery biopsy may show arteritis with giant cells (biopsy proved giant cell arteritis). The syndromes are considered to be different manifestations of giant cell arteritis. Dixon *et al* obtained 10 positive temporal artery biopsy samples in 29 patients with polymyalgia rheumatica.²

The incidence of temporal arteritis, whether diagnosed clinically alone or confined to biopsy proved cases, varies geographically. The disease is almost always confined to white people, and the incidence is higher in Scandinavia and northern Europe (between 17 and 18 cases per 100 000 population aged over 50)^{1 3 4} than in middle France, Spain,¹ and Israel.⁵

The incidence of polymyalgia rheumatica alone is more difficult to determine. Salvarini *et al* studied the incidence of polymyalgia rheumatica in northern Italy and found a figure of 12.7 cases/100 000 population aged over 50.⁶ By contrast, studies from Sweden and Denmark using the same definition of polymyalgia rheumatica obtained figures of 20.4⁷ and 68.3/100 000.¹

Clinical presentation

Giant cell arteritis rarely presents below the age of 50.⁸ It affects many arteries throughout the body, producing symptoms and signs which mimic many other medical and surgical conditions.

Jones has suggested a useful classification of the presenting symptoms of giant cell arteritis⁹:

- (1) *Systemic*—malaise, anorexia, fever, night sweats, weight loss, and depression
- (2) *Myalgic*—proximal, symmetrical muscle pain and stiffness of polymyalgia rheumatica
- (3) *Arteritic*—involvement of the artery may produce:
 - (a) Pain, swelling, erythema, and tenderness over the affected artery
 - (b) Partial occlusion resulting in "claudication-like" symptoms
 - (c) Total occlusion resulting in ischaemia and necrosis of structures supplied by the affected vessel.

Summary points

Polymyalgia rheumatica and temporal arteritis are clinical syndromes affecting elderly people that form part of the spectrum of giant cell arteritis

Both are diseases of people over 50; they occur mainly in white people and show varying incidence across continents

Giant cell arteritis presents in many different forms, which may lead to blindness and stroke

Histological detection of giant cell arteritis remains the only conclusive investigation, supported by a substantially raised erythrocyte sedimentation rate

Diagnosis is largely by exclusion of other conditions, the clinical history, and confirmatory biopsy evidence

Serious consequences can be prevented by rapid treatment with corticosteroids, which should be tailored to the individual patient to avoid side effects in the long term

Nottingham City
Hospital NHS Trust,
Nottingham
NG5 1PB

A J Swannell,
consultant
rheumatologist

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The frequency of clinical features varies considerably in different reports depending on the specialty of the reporting unit, the definition used, the requirement or otherwise for histological confirmation, the use of hospital rather than community studies, and the fact that some patients escape diagnosis. A systemic illness with malaise, anorexia, fever and night sweats, weight loss, and depression is not uncommon. This mode of onset may be confused with infection and malignancy, leading to intensive investigation with the possibility of blindness or stroke developing in the mean time.

The symmetrical proximal muscle pain and stiffness of polymyalgia rheumatica are often associated with giant cell arteritis. There is intense pain and stiffness in the neck, shoulders, and buttocks, which make it impossible for the patient to rise in the morning without rolling out of bed like a log. The stiffness eases during the day. Muscle strength is usually unimpaired

but hindered by pain. If asked, patients often localise the pain to the muscles of the shoulder and neck.

Arteritic involvement by inflammation is most frequently noticed in the superficial temporal arteries, which may stand out and are tender on brushing the hair. There is debate about the value of the state of pulsation of the temporal artery, as atherosclerosis seen on temporal artery biopsy may be responsible for reduced or absent pulsation. Local signs of inflammation may also be found in the posterior auricular, occipital, and facial arteries.¹⁰ Partial occlusion of an artery, producing headache, is a frequent complaint and may be so intense as to cause the patients to sit up in a chair all night. The site of the headache varies considerably and if only temporal pain is accepted for diagnosis some cases will be missed. Jaw pain associated with partial occlusion is the next most common symptom. Arteritis of the maxillary artery produces aching or tiredness in the muscles of the side of the face brought on by chewing and relieved by rest.¹¹ Pain may also be felt in the face and behind the ear and may be associated with vertigo and deafness.

Giant cell arteritis producing problems in the tongue, gums, and throat may cause diagnostic confusion to faciomaxillary surgeons. Lingual artery involvement may cause pain or blanching of the

Ophthalmic features of giant cell arteritis (from Hayreh¹⁴)

- Sudden, painless deterioration of vision in one eye, usually on waking in the morning
- Visual acuity varies from 6/6 to no light perception
- Perimetry shows relative or absolute inferior altitudinal defect, inferior nasal sectoral defect, or central scotoma
- Fundus examination shows optic disc oedema with splinter retinal haemorrhages
- Fluorescein angiography shows filling defects in the optic disc, peripapillary choroidal and choroidal watershed zones, and extensive choroidal filling defects
- Starts as unilateral condition but may become bilateral after days, months, or years

tongue and sometimes impending gangrene.¹² Pain in the gums and teeth has been described, which may suggest a dental origin.¹³

Anterior ischaemic optic neuropathy is the most common and dreaded ocular complication associated with occlusion of the ophthalmic artery, posterior ciliary arteries, choroidal arteries, nutrient arteries of the optic nerve, or central retinal artery. Sudden, painless deterioration of vision in one eye on waking in the morning is the classic presentation. The ophthalmic features have been well reviewed by Hayreh.¹⁴

Giant cell arteritis affecting the vertebrobasilar and sometimes the carotid circulation is well documented at necropsy, but it is difficult to assess how often this results in stroke. The Oxfordshire community stroke project identified eight patients with giant cell arteritis or polymyalgia rheumatica out of 244 cases of new stroke,¹⁵ and it was thought that the proportion of strokes due to giant cell arteritis may be underestimated.

Involvement of the great vessels by giant cell arteritis is often found at necropsy but there is doubt about its prevalence during life, when atherosclerosis is also common in this age group. Giant cell arteritis can result in coronary arteritis and myocardial infarction,¹⁶ but how often this occurs in cardiologists' practice is not known. Aortic incompetence, dissecting aneurysm of the thoracic aorta, and aortic arch syndrome are also well described events that can have serious consequences to which physicians should be alerted.¹⁷

Investigations

Histological detection of giant cell arteritis remains the only diagnostic investigation in polymyalgia rheumatica and temporal arteritis. The artery shows severe intimal thickening and reduction in vessel lumen. The internal elastic lamina is disrupted with fragmentation and sometimes destruction and there is pronounced infiltration by histiocytes, lymphocytes, epithelioid cells, and giant cells in the artery wall, especially the media and intima adjacent to the internal elastic lamina (fig 1). Normal biopsy appearances do not exclude the diagnosis because of possible skip lesions. Temporal artery biopsy samples are positive in 60-80% of patients with giant cell arteritis but in only 15-20% of patients with polymyalgia rheumatica, in whom biopsy for polymyalgia alone is not justified.¹⁸

Fig 1 Top: Low power view of giant cell arteritis showing fragmentation of internal elastic lamina and infiltration by histiocytes, lymphocytes, epithelioid cells, and a giant cell (arrowed). Bottom: High power view of same section showing giant cell in greater detail

The most useful supporting investigation for the clinical diagnosis of polymyalgia rheumatica or giant cell arteritis remains the erythrocyte sedimentation rate or plasma viscosity, despite dispute by some workers.¹⁹ Concentrations of acute phase proteins, especially C reactive protein, are also raised but are not considered more helpful than the erythrocyte sedimentation rate. Increased alkaline phosphatase activity of liver origin occurs in one third to half of patients with both polymyalgia rheumatica and giant cell arteritis. Abnormal tracer uptake has been reported in radionuclide scans with various non-specific abnormalities seen on liver biopsy. Immunological studies on the humoral side give conflicting results, and most evidence favours a cell mediated immune reaction, possibly against an autologous antigen occurring locally in the arteritic lesions of giant cell arteritis.²⁰

Diagnosis

Different criteria for diagnosing giant cell arteritis and polymyalgia rheumatica have been proposed, but clinically the diagnosis of both conditions is largely one of exclusion. There are few if any clinical signs in giant cell arteritis, and the history is thus crucially important. Special attention should be given to asking about weight loss, fever, malaise, morning stiffness, and muscle pains. Any history of headache should be investigated, particularly if it is a new feature and seems to originate outside the skull. Increased prominence and tenderness of the temporal vessels should be sought, together with questions about pain in the jaw, face, gums, or throat. Visual symptoms such as blurring, diplopia, or transient visual loss may give a lead, together with deafness or vertigo.

On examination, arteritis producing tenderness, nodularity, or erythema should be sought in the temporal, posterior auricular, facial, and occipital arteries. Peripheral pulses should be palpated. Blood pressure should be measured on both arms and the presence of bruits sought over the great vessels. The heart should be examined for aortic incompetence. The eyes should be checked for ptosis, orbital swelling, and visual field defects as well as for movement, pupillary reaction, and clarity of the optic discs.

Neurological examination should include the cranial nerves, reflexes, and cerebellar function and a check for nystagmus. The locomotor system should be screened for any pain on movement of the neck, shoulders, or hips. Investigations disclose an increased erythrocyte sedimentation rate in most cases with normochromic anaemia, raised alkaline phosphatase activity, thrombocytosis, and raised IgG concentration. A positive temporal artery biopsy result confirms the diagnosis but a negative result does not exclude it.

Differential diagnosis

At the same time as seeking the above features of giant cell arteritis and polymyalgia rheumatica the clinician must exclude many other probable diagnoses (box). If giant cell arteritis presents with systemic features alone it must be distinguished from infection (pyrexia of unknown origin), malignancy, and myeloma. Anaemia and abnormal liver function values may add to the confusion and cause unnecessary investigation.

Polymyalgia rheumatica can usually be distinguished from rheumatoid arthritis by the absence of peripheral synovitis²¹ but is possibly the commonest cause of confusion in elderly patients. In polymyositis the predominant feature is weakness rather than the intense pain of polymyalgia rheumatica. Screening of plasma protein values and a chest x ray film may also help to avoid confusion with myeloma and miliary tubercle.

Differential diagnoses

Polymyalgia rheumatica

- Neoplastic disease
- Cervical spondylosis
- Rheumatoid arthritis
- Connective tissue disease
- Myeloma
- Leukaemia
- Bone disease (osteomyelitis)
- Hypothyroidism
- Miliary tubercle

Temporal arteritis

- Dental conditions
- Trigeminal neuralgia or sinus disease
- Otological conditions
- Retinal vascular accident
- Other causes of ophthalmoplegia

Pain in the jaw may suggest dental causes and pain in the face trigeminal neuralgia or sinus disease. Pain in the ear with or without vertigo raises the possibility of otological conditions. Visual loss may be ascribed to a retinal vascular accident. The ophthalmoplegia of giant cell arteritis varies in severity and which muscle it affects, unlike other types of ophthalmoplegia.⁹

Sometimes the diagnosis of giant cell arteritis and polymyalgia rheumatica may seem clear cut, but quite often it pays to perform screening tests for other differential diagnoses when the diagnosis is less obvious. After starting steroids other diagnoses may be masked or take on a more rapid course with dangerous consequences.

Management

Temporal artery biopsy is not justified in a straightforward case of polymyalgia rheumatica with no sign of giant cell arteritis, as blindness is uncommon in these patients.²² The question of temporal artery biopsy in giant cell arteritis and temporal arteritis is still debated. A classic presentation of temporal arteritis with high erythrocyte sedimentation rate may make this investigation seem unnecessary, but such presentations are not common and patients require long term steroids with the potentially serious side effects.

Temporal artery biopsy not only confirms the diagnosis but excludes other systemic vasculitides such as polyarteritis nodosa and Wegener's granulomatosis.²³ There may be delay in arranging a biopsy but this need not delay steroid treatment. Treatment will lessen the chance of a positive biopsy result, but positive results after starting steroids have been reported up to a week²⁴ or even up to three and six months in some cases.²⁵ Positive temporal artery biopsy findings at the initiation of treatment give the physician confidence to continue steroid treatment which may have side effects and avoid possible litigation.¹⁴

Corticosteroids are essential for the treatment of polymyalgia rheumatica and temporal arteritis, as they rapidly relieve the incapacitating symptoms and reduce the incidence of blindness.²⁶ The use of non-steroidal anti-inflammatory drugs has been advocated from the United States,²⁷ but most physicians find steroids are necessary to achieve complete control of symptoms. The response to steroids is dramatic, with relief of symptoms in 48-72 hours and producing very grateful patients. The beneficial effects of treatment must be balanced against the unwanted side effects.

There are few prospective studies of the correct starting dose, but the conclusion is that 40 mg prednisolone for giant cell arteritis and 15 mg prednisolone for polymyalgia rheumatica are appropriate initial treatments for most patients.²⁶ A recent study in general practice suggested that the initial steroid dose used in patients with polymyalgia rheumatica is still generally too high.²⁸ Some ophthalmologists recommend 80 mg prednisolone for giant cell arteritis because of the higher risk of arteritic complications. Megadose intravenous corticosteroid treatment has been suggested when patients with arteritis have visual signs or symptoms such as amaurosis fugax or early evidence of second eye involvement.¹⁴

There is little information on the rate of corticosteroid reduction once initial symptoms are controlled, but gradual reduction by weekly decrements of 5 mg, titrating the treatment alterations against the clinical picture rather than the laboratory results, has been recommended.²² Once 10 mg is reached it is suggested that 1 mg every two to four weeks is sufficient. At that stage my policy is to review patients at monthly intervals, titrating the dose of prednisolone against the symptoms and the erythrocyte sedimentation rate. Reduction schedules can only be suggestions, as cases vary greatly and must be judged individually. Regular review of these patients is important.

Relapses are more likely during the initial 18 months of treatment and within one year of withdrawal of steroids. Patients should be asked to report back urgently if arteritic symptoms occur. No way of predicting those who will relapse has been discovered, but arteritic relapse in patients who presented as cases of polymyalgia rheumatica is unusual.

Most studies indicate that between one third and one half of patients can stop steroids after two years. The risk of relapse must be balanced against the steroid related side effects.²⁹⁻³⁰ Fractures and severe infections are the most common complications and seem dose related. Recent work suggests that intermittent cyclical etidronate may prevent some of the corticosteroid induced bone loss in patients with temporal arteritis receiving high dose steroids if given when treatment is begun.³¹ In patients in whom steroid reduction or withdrawal is difficult azathioprine has been used. In the later stages of steroid reduction adding non-steroidal anti-inflammatory drugs may help the pseudorheumatism that often develops.²⁶ Methotrexate has been shown to have a modest steroid sparing effect,³² though not in every case.³³

Polymyalgia rheumatica and temporal arteritis are among the most rewarding diseases for a clinician to diagnose and treat because the unpleasant symptoms and serious consequences can be rapidly prevented with corticosteroids. Objective means of determining

the prognosis in individual patients and decisions concerning the duration of treatment remain empirical and require careful supervision.

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