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Scleritis and temporal arteritis

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Summary

Thirty consecutive patients with severe scleritis or episcleritis were admitted as in-patients to the Medical Ophthalmology Unit and assessed for systemic disease. There were seventeen women and thirteen men. The mean age was 53 with a median of 57 (range 23–83). Eighteen of the patients had scleritis: eleven of these had evidence of connective tissue disease and three of them had temporal arteritis. Twelve patients had episcleritis: six of them had a collagen disease and one of them developed temporal arteritis. This high incidence of temporal arteritis in association with scleritis has not been previously reported. It is important to diagnose and treat overt temporal arteritis early with parenteral steroids so that ischaemic papillopathy can be avoided.

A higher incidence of collagen diseases than previously described is reported in episcleritis. It is thought that this is secondary to selection since patients with the usual self-limiting episcleritis are not normally referred for further in-patient investigation.

In no patient was more than one significant diagnosis made. There was no significant medical illness in only 11% of patients with scleritis and 33% of patients with episcleritis. The majority of the non-collagen diseases (e.g. hypertension) were not previously recognized. In none of the patients with temporal arteritis was the diagnosis made before admission. It is concluded that full examination and investigation for underlying disease is indicated in both scleritis and severe episcleritis.

Introduction

Scleritis is well known as being associated with rheumatoid arthritis and less commonly with polyarteritis nodosa, tuberculosis, syphilis, Wegener's granulomatosis, lupus erythematosus and herpes zoster. Other diseases such as ankylosing spondylitis, Behçet's disease, dermatomyositis, polychondritis and temporal arteritis are also described. The incidence of such diseases in scleritis has been assessed at about 50% (Watson, 1966; Lyne and Pitkeathly, 1968; Lyne, 1974). Similar conditions are described in episcleritis and the incidence is usually assessed at about 10%. No case of temporal arteritis in association with scleritis or episcleritis is to be found in the British or American literature of the last 25 years. A review of the ocular manifestations of eighty-four cases of temporal arteritis revealed no cases of scleritis or episcleritis (Bruce, 1949).

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Patients and methods

Thirty consecutive patients with severe scleritis or episcleritis who were admitted as in-patients to the Medical Ophthalmology Unit at Lambeth Hospital were assessed for underlying systemic disease. All the patients presented to ophthalmic clinics primarily with eye symptoms; on admission, eight patients were known to have underlying rheumatoid arthritis and the cases of migraine, bronchiectasis and coronary artery disease were already diagnosed. A full medical and ophthalmological history was taken from each patient and all the patients were examined by both an ophthalmic surgeon and a physician with a special interest in inflammatory ocular disease. Routine investigations included a full blood count and erythrocyte sedimentation rate (ESR); radiography of the chest, lumbar spine, sacro-iliac joints, hands and feet; urea and electrolytes, creatinine clearance, liver function tests including protein electrophoresis and serum immunoglobulins; antinuclear factor and sheep cell agglutination test; microscopy and culture of midstream specimen of urine and Wassermann and *Treponema pallidum* haemagglutination tests.

Results

Of the eighteen patients with scleritis, eleven were women and seven were men. The mean age was 55. The disease was bilateral in seven patients and

unilateral in eleven. Three of the patients with rheumatoid arthritis had unilateral scleromalacia perforans. Eight patients had underlying rheumatoid arthritis, three temporal arteritis, two allergy and one each an arthropathy of unknown cause, bronchiectasis and coronary artery disease. In two patients no underlying pathology was found (Table 1). All the patients who were diagnosed as having rheumatoid arthritis fulfilled the criteria of the American Rheumatism Association for 'classical' or 'definite' rheumatoid arthritis. Seven of the rheumatoid patients were sheep cell agglutination seropositive and in only one of the eight was the ESR less than 25 mm in 1 hr. In six of the rheumatoid patients there were radiological changes of the joints. One patient had subcutaneous nodules. The two patients with allergic scleritis had a long history of abnormal hypersensitivity and were both female and young (aged 26 and 35).

TABLE 1. Systemic diseases in eighteen patients with scleritis

	No.	%
Rheumatoid arthritis	8	44
Temporal arteritis	3	16
Allergy	2	11
Arthropathy (? cause)	1	6
Bronchiectasis	1	6
Coronary artery disease	1	11
Nil	2	11
Total	18	100

In the episcleritis group of twelve patients, the sex ratio was equal. The mean age was 45. The episcleritis was bilateral in seven patients and unilateral in five. Two of the patients had arthropathy of unknown cause, two had hypertension, two had rheumatoid arthritis and one each had migraine and temporal arteritis. In four patients no underlying disease was found (Table 2). Of the two patients with arthropathy of unknown cause one, a woman of 26, had radiologically obliterated sacroiliac joints and was thought to have mild ankylosing spondylitis. The other, a man of 40, had had large joint arthralgias associated with jaundice: this was thought to be type A hepatitis. The episcleritis started at the same time as the jaundice but persisted 3 months after the liver function tests had returned to normal. Both the hypertensives were newly diagnosed and needed hypotensive therapy. Both the rheumatoid patients were seropositive and had radiological joint changes; one had an associated pleural effusion. The patient with migraine had late onset cluster headaches but no clinical evidence of temporal arteritis.

TABLE 2. Systemic diseases in twelve patients with episcleritis

	No.	%
Arthropathy (? cause)	2	17
Hypertension	2	17
Rheumatoid arthritis	2	17
Migraine	1	8
Temporal arteritis	1	8
Nil	4	33
Total	12	100

Case histories

Case 1

A 60-year-old man was admitted with a 9-day history of severe bifrontal and bitemporal headache, painful red eyes and photophobia. The headache was constant and throbbing and prevented him sleeping. He had noticed blurring of vision in the inferior field of his left eye. He was also nauseated and anorexic. On examination he had palpable tender temporal arteries which were weakly pulsatile. Episcleral and scleral vessels were congested on the left but not on the right. The left visual field showed an inferior scotoma suggesting an ischaemic papillopathy. The ESR was 108 mm in 1 hr. There was a neutrophil leucocytosis and a rise in α_2-globulins. A clinical diagnosis of temporal arteritis with left ischaemic papillopathy and scleritis was made. He was started on i.m. hydrocortisone. Three days later he had become asymptomatic and was started on oral prednisolone. After 10 days, he felt well, the sclerae were normal and the ESR was 17. He was followed-up in out-patients where symptoms and ESR were monitored. After 18 months the prednisolone was stopped. He remains symptom-free.

Case 2

A 68-year-old woman was admitted with bilateral painful red eyes. She had first developed symptoms 6 weeks previously and she had had three subsequent relapses each lasting a few days. She also had recurrent severe bitemporal headaches. This was associated with anorexia and a 12·7 kg weight-loss. She was taking thyroxine 0·2 mg daily for hypothyroidism. On examination she was thin and had bilateral scleritis. The ESR was 98 and there were increased α_2- and γ-globulins. A temporal artery biopsy showed a giant cell arteritis. A diagnosis of temporal arteritis was made. She was treated with local and systemic steroids. Her ESR was 37 after 3 weeks' therapy. Three years later she remains on prednisolone 10 mg daily because of recurrent scleritis.

Case 3

An 83-year-old woman was admitted with a 3-month history of red painful watery eyes and poor vision in the right eye. This was associated with a severe bilateral headache in the frontal and temporal areas. On examination she was a thin, partially demented old lady with bilateral scleritis, keratitis and iritis. Investigations revealed a neutrophil leucocytosis, an ESR of 29 and an increased plasma γ-globulin. Temporal artery biopsy showed fragmentation of the internal and external elastic laminae, fibroelastic intimal thickening and an inflammatory reaction composed mainly of lymphocytes in the adventitia: this was interpreted as temporal arteritis. She was treated with oral prednisolone and neomycin and atropine eye drops. On this her headache resolved, and the eyes greatly improved. She was discharged to geriatric care on prednisolone 30 mg daily.

Case 4

A 53-year-old man developed bilateral episcleritis which was treated at another hospital with oral oxyphenbutazone and local steroids. Four months after the onset of episcleritis he developed bitemporal and bifrontal throbbing headaches, a right relative inferior scotoma and general malaise. On examination the temporal arteries were mildly tender, there was a mild red-green colour defect in the right eye and subjectively the vision of the right eye was worth 80% of that of the left. The fundi and ophthalmodynamometry were normal. Investigations showed an ESR of 85, a neutrophil leucocytosis, a raised α_2-globulin and an alkaline phosphatase of 18 KAu/100 ml. A clinical diagnosis of temporal arteritis was made and on prednisolone 40 mg/day he became asymptomatic apart from the visual field defect and the investigations returned to normal. The prednisolone was gradually reduced. At a dose of 7·5 mg/day, he once again developed a headache and the ESR was found to be 60. One year after the introduction of prednisolone he still needs 10 mg/day to prevent symptomatic and haematological relapse.

Discussion

Scleritis is well recognized as a complication of rheumatoid arthritis and is usually associated with an exacerbation of the generalized disease. The majority respond to systemic and local corticosteroids but some seem to require the addition of immunosuppressive therapy (Jayson and Jones, 1971). The rare complication of scleromalacia perforans has a poor prognosis for vision (Williams and Rosenthal, 1959).

The main finding of this study is a 16% incidence of temporal arteritis in scleritis and an 8% incidence in episcleritis. The disease is almost unreported under the age of 50 (Fauchald, Rygvold and Oyestese, 1972). The incidence among the susceptible age group is therefore higher than this. The diagnosis is essentially clinical on the history of headache, tender temporal arteries, jaw pain, difficulty in opening the mouth and inflamed scalp with or without cutaneous nodules. Confusion or depression may predominate. Symptoms of polymyalgia may be present and there is often anorexia and weight-loss. There is often a normocytic anaemia, a neutrophil leucocytosis, a raised ESR and an increase in α-globulins (Hamilton, Shelley and Tumulty, 1971; Turner *et al.*, 1974). Temporal artery biopsy is often helpful in confirming the diagnosis but false negative results may occur because of skip lesions (Birkhead, Wagener and Shick, 1957). Raised hepatic alkaline phosphatase has recently been reported in fifteen cases (Hall and Hargreaves, 1972). An association with thyroid disease has been claimed (Thomas and Croft, 1974). Treatment with anti-inflammatory drugs such as aspirin gives some symptomatic relief but over 30% of patients progress to visual failure; the introduction of high dose cortosteroids rapidly alleviates symptoms and is associated with the prevention of ischaemic retinal problems (Ross Russell, 1959). It has been claimed that oxyphenbutazone is effective in episcleritis (Watson *et al.*, 1966). However, if there is clinical suspicion of temporal arteritis, high dose systemic steroids are immediately indicated.

In a previous study from this department on uveitis it was concluded that, in view of the poor return, widely cast investigative routines were not indicated (James *et al.*, 1969). It seems that an investigative routine is more fruitful in scleritis and severe episcleritis. The following is recommended:

(a) General medical history and examination with a particular search for arthritis or a collagen disorder.

(b) Rheumatoid factor.

(c) DNA binding capacity and serum complement for systemic lupus.

(d) X-rays of hands and sacro-iliac joints.

(e) ESR to monitor progress and response to treatment.

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Notes