SCIENTIFIC CORRESPONDENCE

Orbital myositis in scleritis

Z F H M Boonman, R J W de Keizer, H S Graniewski-Wijnands, P G Watson

Br J Ophthalmol 2003;**87**:38–42

Aims: To investigate the association between scleritis and myositis.
Methods: Retrospective, non-comparative case series. Records and ultrasonograms were examined of 132 patients, with a diagnosis of episcleritis or scleritis, who attended the ophthalmology department at Leiden University Medical Center between 1997 and 2000. 103 were eligible for comprehensive examination. Medical records were evaluated. Ultrasonography was performed in all patients diagnosed with episcleritis or scleritis. Clinical features, precipitating factors, systemic associations, ocular complications, treatment, and outcome of each patient were assessed.
Results: Of the 103 patients, 27 (26.2%) had episcleritis and 76 (73.8%) had scleritis. Myositis was found to be present in 11 patients. It was present in 14.5% of all patients with scleritis and 30.5% of those in whom the posterior sclera was affected. The presence of the associated myositis did not worsen the visual prognosis and the presence of myositis was not associated with other systemic diseases. There were no cases of unilateral scleritis with bilateral orbital myositis. During an attack ocular complications were more common in patients with scleritis and myositis (64%) than in patients with scleritis alone (30.4%), indicating a more diffuse and potentially dangerous inflammation. There was no evidence that the inflammatory changes in the orbit had spread to involve the sclera, so it is assumed that the muscle changes are an extension of a generalised response to intense inflammation of the episclera and sclera.
Conclusion: This study found a frequent association between myositis and scleritis. Prognosis for vision was not affected by coexistence of myositis.

Inflammation of the wall of the eye and its mesodermal coverings can present in many ways from the benign simple episcleritis to the severe necrotising scleritis and is sometimes known to involve the overlying muscles. The differential diagnosis of episcleritis from scleritis and between diffuse, nodular, and necrotising disease affecting the anterior segment of the eye is well recognised.[1] It is also known that the condition with which the patient first presents usually continues throughout the course of the disease.[2] Scleritis is a serious, usually painful, progressive disease of the sclera itself, which if left untreated can seriously affect vision. Diffuse and nodular anterior scleritis if treated early does not normally result in any major complications but if the inflammation extends to or affects the posterior segment of the eye, it frequently results in visual loss. Recent studies using B scan ultrasonography have shown that both episcleritis and scleritis may involve the posterior segment even though there are no clinical signs. Even minor changes in the posterior segment can threaten vision.[3]

Primary idiopathic orbital myositis is a relatively common subtype of idiopathic orbital inflammatory disease (IOID) in which one or more of the extraocular muscles are affected. The clinical course of orbital myositis is usually characterised by an acute onset, severe pain in and around the eye, pain on movement, occasional diplopia and chemosis, a rapid response to systemic steroids and short duration, although recurrences may occur in some cases.[4 5] These symptoms and signs are very similar to those encountered in patients with posterior scleritis.

This study was instituted to determine the importance of the association between myositis and scleritis, whether the extraocular muscles were involved secondary to the scleral disease, and whether it was sometimes bilateral even though only one eye was affected with the scleral disease.

MATERIALS AND METHODS

We reviewed the records of 132 patients with episcleritis or scleritis attending the department of ophthalmology at the Leiden University Medical Center between January 1997 and April 2000. Of the 132 records identified, 29 were excluded because of insufficient clinical detail to ensure a correct diagnosis or lack of insufficient follow up data for the 3 year study period. The follow up period was at least 6 months. All the patients were examined during the 3 years before the end of the study.

Scleritis was divided into anterior and posterior. Anterior scleritis incorporated diffuse, nodular, necrotising with inflammation (necrotising), and necrotising without inflammation (scleromalacia perforans). Episcleritis was not further subclassified into diffuse or nodular.[1]

In this department all patients diagnosed with scleritis or episcleritis have B-scan ultrasonographs in addition to the clinical and laboratory examinations. The diagnosis of myositis was made clinically and by ultrasonography using the techniques followed in the orbital clinics.[6 7] Ultrasonically myositis shows diffuse thickening of both the tendon and muscle fibres with low internal reflectivity. One person (HGW), who is experienced in making both A-mode and B-mode ultrasounds of the orbit, performed all the patients' scanning, using the Biovision-B-S with a 10 MHZ transducer and a standardised A-mode transducer and techniques of standardised ultrasonography. The transducer was placed perpendicular to the medial rectus muscle so that the muscle's thickness could be measured accurately. Other extraocular muscles were evaluated for oedema, fibre structure, and cysts; however, the thickness could not be quantified since the echo's transducer could not always be placed in an angle of 90° towards the measured muscle. In this clinic the normal upper diameter limit of a medial rectus is 5.4 mm measured over 45 patients. All the patients were examined by an internist, rheumatologist, otorhinolaryngologist, or neurologist. Patients diagnosed with Graves' disease were excluded from the study. The visual acuity was measured at presentation and at the end of follow up. Visual loss was considered to be defective if there was a reduction of two Snellen lines or more. Another disease or complication which may have caused a reduction in visual acuity was recorded to determine that the visual loss could be solely attributed to the scleral inflammation. The data

Table 1 General characteristics of patients with episcleritis and various scleritis types

Characteristic	Episcleritis	Scleritis				Total
		Diffuse anterior	Nodular anterior	Necrotising	Posterior	
Patients (%)	27 (26.2)	19 (18.4)	16 (15.5)	5 (4.9)	36 (34.6)	103 (100)
Eyes	29	26	22	8	48	133
General features (n=103)						
Age, mean (SD)	42.6 (13.3)	42.5 (14.4)	51.1 (15.3)	44.0 (14.8)	53.7 (17.9)	47.9 (16.2)
Women (%)	18 (66.7)	14 (73.7)	13 (81.3)	3 (60.0)	20 (55.6)	68 (66.0)
Age: women mean (SD)	42.3 (14.7)	42.5 (10.5)	50.1 (14.7)	48.0 (19.1)	55.6 (19.5)	48.0 (16.4)
Men (%)	9 (33.3)	5 (26.3)	3 (18.75)	2 (40.0)	16 (44.4)	35 (34.0)
Age: men mean (SD)	43.3 (10.5)	42.6 (24.0)	55.3 (20.7)	38 (5.7)	51.4 (15.9)	47.7 (16.1)
Bilateral	2 (7.4)	7 (36.8)	6 (33.3)	3 (60)	12 (33.3)	30 (29.1)
Scleritis patients without myositis	27 (29.3)	19 (19.6)	16 (16.3)	5 (5.4)	25 (27.2)	92 (100)
Visual loss at presentation (%)	2 (7.4)	2 (5.6)	5 (31.3)	3 (60)	11 (11.9)	23 (25)
Visual loss after treatment (%)	–	–	5 (31.3)	1 (20)	9 (36)	15 (16.3)
Associated eye abnormalities						
Cornea abnormalities %	2 (7.4)	–	3 (18.8)	–	1 (4)	6 (6.5)
Eye diseases (uveitis, retinal detachment, glaucoma and cataract) %	1 (3.7)	–	5 (31.3)	4 (80)	12 (48)	22 (23.9)

N=103.
SD=standard deviation.

concerning the patients were entered into a computerised database to ensure that they were retrievable. Forward stepwise logistic regression was undertaken to determine whether any of the recorded variables had predictive value for responsiveness to treatment, outcome in terms of visual loss, and the presence of complications. The SPSS statistical software version 10 (SPSS Inc) was used for statistical analyses.

RESULTS

Of the 103 patients, 76 had scleritis and 27 had episcleritis (Table 1). Myositis was found to be present in two patients with posterior scleritis, eight patients with both anterior and posterior scleritis, and in one patient with anterior episcleritis and posterior scleritis (Table 2). The eight patients with anterior scleritis had, besides their myositis and posterior scleritis, diffuse anterior scleritis (six) and nodular anterior scleritis (two).

There were five females and six males with both scleritis and myositis whose ages were 27–77 years with a mean age of 51.9 years (SD 13.4 years).

Thirty patients had bilateral involvement, including two with episcleritis and 28 with scleritis. Four patients with both scleritis and myositis had scleral disease in both eyes (Table 2). One patient with bilateral episcleritis and posterior scleritis in the left eye had myositis in the left eye only. There were no patients who had scleritis in one eye and myositis in both eyes. Three patients with bilateral scleritis also had myositis in both eyes. Thus myositis only occurred in an eye with posterior scleral inflammation. Clinically, those with scleritis and myositis had periocular pain, pain on movement (90.9%), proptosis, diplopia, headache, chemosis, red eyes, and swollen eyelids (Table 2). Patients with the combination scleritis and myositis had severe pain on ocular movement (OR = 26.8: 3.3 to 220.3), whereas in the episcleritis and scleritis group without myositis only 25 patients (26.4%) had pain on movement, 44% of the patients with posterior scleritis also had pain on movement. There was only one patient in whom the B-scan abnormality did not correlate with the clinical diagnosis of myositis. The medial rectus muscle, was predominantly affected.

The visual acuity was measured at presentation and at the end of follow up in 101 patients. There were three patients with both scleritis and myositis who had loss in visual acuity both at presentation and after treatment (Table 2). One patient had a serous retinal detachment in the affected eye with scleritis and developed a cataract in the same eye. Another patient with unilateral disease had a concurrent iridocyclitis and developing cataract. The last patient with bilateral scleritis and myositis had bilateral concomitant secondary glaucoma. In the scleritis group without myositis, 21 patients had visual loss at presentation of whom 15 still had a decrease in visual acuity after treatment. Vision was especially affected in patients with exclusively posterior scleritis both at presentation (44%) and after treatment (36%). Patients with both scleritis and myositis did not have any greater risk of developing deficient vision at presentation and after treatment compared to patients with scleritis alone. Ocular complications were more common in those patients with scleritis and myositis (64%), consisting of cataract, keratitis, high intraocular pressure, glaucoma, and uveitis compared to those who had scleritis (30.4%) alone (OR = 4.0: 1.1 to 14.8). Patients were treated both for their scleritis and additional ocular complication. The additional eye disease responded completely after treatment.

One third of the patients with scleritis and one in 10 of those with episcleritis had an associated systemic disease (Table 3) but only two of the 11 patients with both scleritis and myositis had an associated systemic disease. Patients with an associated systemic disease had a greater risk of developing the ocular complications of scleritis after their remission (OR = 3.7: 1.4 to 9.7).

Response to treatment in patients with both scleritis and myositis was not worse statistically compared to the group of patients with only scleritis. All patients with both scleritis and myositis went into complete remission after treatment. Some of these patients were occasionally troubled with pain and redness or developed a recurrence. However, they did not have more recurrences or ocular complications after their remission when compared with the scleritis group.

DISCUSSION

Scleritis has been known to be associated with swelling of the adjacent muscles and their fascial sheaths for many years.[8–10] However, the significance and the frequency of this association has not yet been fully explored.

The inflammation of the muscles and their sheaths is poorly defined and has been termed orbital myositis, extraocular muscle fasciitis, and/or tenonitis. The muscles and sheaths have been found to be infiltrated by inflammatory cells and the inflammation may be accompanied by orbital granulomas similar histologically to that found in scleral disease and in some instances contiguous with it. Both orbital myositis and scleritis can be extremely painful and both tissues can be

Table 2 Characteristics of patients with both myositis and scleritis

Characteristic	Posterior scleritis and anterior scleritis/ episcleritis	Posterior scleritis	Total
Patients with only scleritis	30 (29.1)	6 (5.8)	36 (34.9)
Patients with myositis and scleritis	9	2	11 (100)
Eyes	12	3	15
General features (n=11)			
Age, mean (SD)	54.4 (11.8)	40.5 (19.1)	51.9 (13.4)
Women	4	1	5 (45.5)
Age: women mean (SD)	55.0 (15.6)	54.0	54.8 (13.5)
Men	5	1	6 (54.5)
Age: men mean (SD)	54.0 (9.9)	27.0	49.5 (14.2)
Bilateral scleritis (%)	3	1	4 (36.4)
Bilateral myositis (%)	2	1	3 (27.3)
Pain and other symptoms			
Foreign body feeling (%)	1	–	1 (9.1)
Medium pain (%)	3	–	3 (27.3)
Severe pain (%)	5	2	7 (63.4)
Pain on movement (%)	8	2	10 (90.9)
Periocular pain (%)	7	2	9 (81.8)
Headache (%)	4	2	6 (54.5)
Red eyes (%)	9	–	9 (81.8)
Swollen eyelids (%)	1	–	1 (9.1)
Chemosis (%)	3	1	4 (36.4)
Proptosis (%)	3	2	5 (45.6)
Impaired ocular movement (%)	3	2	5 (45.6)
Diplopia (%)	2	–	2 (18.2)
B-Ultrasonography			
Muscle abnormalities (%)	9	2	11 (100)
Sclera abnormalities (%)	9	2	11 (100)
B-ultrasound comparable with clinical myositis symptoms (%)	8	2	10 (90.1)
Ocular complications			
Visual loss at presentation (%)	3	–	3 (27.3)
Recurrences (%)	3	2	5 (45.5)
Sclera melting (%)	1	–	1 (9.9)
Associated eye abnormalities			
Cornea abnormalities (%)	2	–	2 (18.2)
Eye diseases (uveitis, retinal detachment, glaucoma and cataract) (%)	4	1	5 (45.5)
Responsiveness to treatment			
Poor response (%)	4	–	4 (36.4)
Good response (%)	5	2	7 (54.5)
Outcome			
Complete remission (%)	4	1	5 (45.5)
Remission with ocular complications (%)	5	1	6 (54.5)
No remission (%)	–	–	–
Visual loss after treatment (%)	3	–	3 (27.3)

N=11.
SD=standard deviation.

involved in systemic diseases particularly those which have a systemic vasculitis. Idiopathic orbital myositis is currently included within idiopathic orbital inflammatory syndromes.[4 11] It is rarely associated with any other condition and differs from dysthyroid eye disease (Graves' orbitopathy).[4 11] As a consequence any patient with Graves' disease was excluded from this study, except one patient with anterior episcleritis who had thyrotoxicosis without muscular involvement.

In this series all patients who presented with any form of episcleral or scleral inflammation had a B-scan ultrasonography, which was performed by the same individual. This B-scan ultrasound included an orbital scan of the ocular muscles and their sheaths.[6 7 12] We consider that scleritis should be added to the list of conditions normally associated with myositis because orbital myositis was found frequently enough in both patients with posterior scleritis alone (30.5%) or in those who had both anterior and posterior segments (25%) involved (Figs 1 and 2).[11] In this study all patients with myositis had posterior scleritis. Of these nine had involvement of the anterior sclera as well. They all had the clinical symptoms of orbital myositis and had at least one thickened medial rectus on B-mode ultrasound. The pain was often unbearable. All patients consulted a general doctor or ophthalmologist within 2–5 days after the first symptoms because of the severity of the disease. Pain on movement occurred in patients with scleritis alone (26.4%), although not as frequently and severe as in patients with both scleritis and myositis (90.9%). Therefore, this symptom is very indicative of the combination of scleritis and myositis. Patients with both scleritis and myositis had more additional ocular problems than patients with scleritis alone, indicating a more widespread and potentially more damaging inflammatory response (OR = 4.0: 1.1 to 14.8). There were only two patients with scleritis and myositis who had an associated disease. There is no suggestion that orbital vasculitis, pseudotumour, rheumatic arthritis, or other connective tissue diseases are related to this combined condition.

The average age of presentation of patients with scleritis and myositis was 51.9 years (SD 13.4 years). This is much older than those who present with a primary orbital myositis who are normally young to middle aged with a mean age of 36.8 years (SD 17.7 years).[13–15]

Table 3 Associated diseases in scleritis and episcleritis

Systemic disease	Scleritis	Scleritis + myositis	Episcleritis
Rheumatic arthritis	4		1
Sjögren's syndrome	1		
Wegener's granulomatosis	4		
Periarteritis nodosa	2		
Local periarteritis nodosa	1		
Reiter syndrome	1		
Systemic vasculitis	1		
Systemic lupus erythematosus	4		
Cogan syndrome	1	1	
Ankylosing spondylitis	3		
Crohn's disease		1	1
Sarcoidosis	1		
Thyrotoxicosis			1
Rhinosporidiosis	1		
Total (n=103)	24 (31.5%)	2 (2.7%)	3 (11.1%)

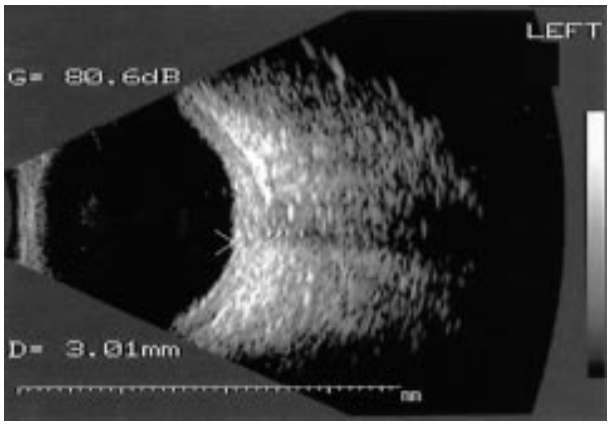

Figure 1 B-mode ultrasound before treatment of a 75 year old female patient with scleritis and myositis in both eyes. The patient's left eye has been made visible on ultrasound. The sclera is thickened with 3.01 mm. D= distance.

Other studies into orbital myositis show frequent involvement of the horizontal recti, although every muscle may be engorged in this condition.[16 17] In a large series of patients it was shown that the frequency of muscle involvement, in decreasing order, was medial rectus (43%), superior rectus (19%), lateral rectus (17%), superior oblique (9%), inferior rectus (7%), and inferior oblique (5%).[17] Although unilateral, single muscle inflammation with tendon association is most common, orbital myositis may be bilateral, in which case one or more extraocular muscles with or without their tendons may be involved.[17–19] It has not been possible to determine in this series whether the swelling of the muscles seen ultrasonographically was a myositis or just swelling of the fascia. It is probable that the swelling of the muscle and its sheath is secondary to the scleral inflammation rather than the inflammatory response extending into the scleral coats from a primary myositis, because there was no evidence of other orbital disease in these patients. The age group of the patients is that of scleritis rather than myositis and, in addition, myositis alone was never present in the opposite orbit to the eye which had scleritis or the scleritis-myositis combination.

Scleritis, whether anterior or posterior, is accompanied by oedema of the overlying episclera but high quality ultrasonography shows that the posterior episclera and the muscle sheaths can be involved in an inflammatory process without the sclera apparently being involved. If episcleral inflammation affects the extension of the episclera onto the optic nerve, then vision can be affected and, if it tracks forward on and around the muscle, it can present as an episcleritis as occurred in one patient in this series.

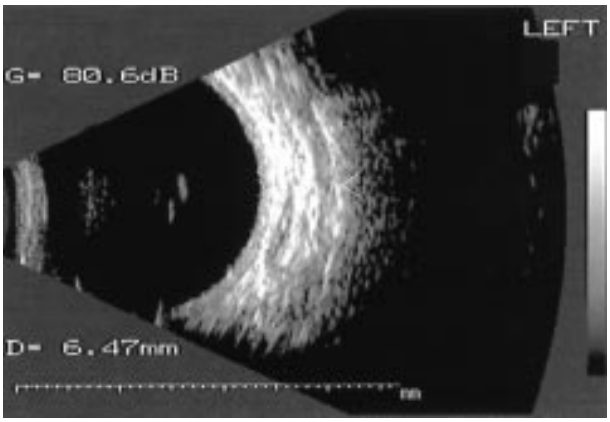

Figure 2 B-mode ultrasound before treatment of the same patient as in Figure 1. The patient's left eye has been made visible on ultrasound. The medial rectus muscle is thickened (6.47 mm) and inflamed. D= distance.

No complications directly related to the myositis could be identified but the source of the very severe pain and pain on movement experienced by patients with the combination of scleritis and myositis was almost certainly due to inflammation of the muscles rather than derived predominantly from the sclera. The treatment of myositis and scleritis is identical and both responded to non-steroidal anti-inflammatory agents or systemic steroids. Additional immunosuppression therapy was never required. As anticipated the myositis recovered rapidly but there was a delay in the final resolution of the scleral inflammation. B-scan ultrasound of the orbit should be included in the investigation of scleral disease as the presence of a myositis indicates a more extensive disease and, as a consequence, more aggressive therapy may be indicated.

Financial support: There was no particular funding involved for this study.

Commercial interest: None.

Authors' affiliations

Z F H M Boonman, R J W de Keizer, H S Graniewski-Wijnands, P G Watson, Department of Ophthalmology, Leiden University Medical Center, Leiden, Netherlands

P G Watson, Department of Clinical Ophthalmology, Institute of Ophthalmology and Moorfields Eye Hospital, London, and Addenbrooke's Hospital, Cambridge, UK

Correspondence to: Zita F H M Boonman, Leiden University Medical Center, Department of Ophthalmology (J3-S), PO Box 9600, 2300 RC, Leiden, Netherlands

Accepted for publication 29 July 2002

REFERENCES

1 **Watson PG**, Hayreh SS. Scleritis and episcleritis. *Br J Ophthalmol* 1976;**60**:163–91.
2 **Tuft SJ**, Watson PG. Progression of scleral disease. *Ophthalmology* 1991;**98**:467–71.
3 **McCluskey PJ**, Watson PG, Lightman S, *et al*. Posterior scleritis: clinical features, systemic associations, and outcome in a large series of patients. *Ophthalmology* 1999;**106**:2380–6.
4 **Keizer de RJW**. Idiopathic orbital inflammation and vasculitis orbitae. In: Keizer de RJW, Jager MJ, eds. *Handbook of ocular immunology*. Buren: Aeolus Press, 1998:227–45.
5 **Watson PG**, Hazleman BL. The sclera and systemic disorders. *Major problems in ophthalmology*. Vol 2. London, Philadelphia: Saunders, 1976.

6 **Frazier Byrne S**, Green RL. Orbital echography. In: Tasman W, Jaeger EA, eds. *Duane's clinical ophthalmology*. Philadelphia: JB Lippincott, 1993;v 2, chapter 26.
7 **Byrne SF**. Standardized echography of the eye and orbit. *Neuroradiology* 1986;**28**:618–40.
8 **Birsch-Hirschfeld A**. Zur pathologie und therapie der orbitalphlegmonie hochgradige verbiegung und lakunare faserdegeneration im optikus. *Albrecht v Graefes Archiv fur Ophthalmol* 1910;**71**:333–49.
9 **Derby GS**, Verhoeff FH. Massive granuloma of sclera (brawny scleritis) with a report of an unusual case. *Arch Ophthalmol* 1916;**45**:20–32.
10 **Bertelsen TI**. Acute sclerotenonitis and ocular myositis complicated by papillitis, retinal detachment and glaucoma. *Acta Ophthalmol* 1960;**38**:136–52.
11 **Lacey B**, Chang W, Rootman J. Nonthyroid causes of extraocular muscle disease. *Surv Ophthalmol* 1999;**44**:187–213.
12 **Ossoinig KC**. Standardized echography: basic principles, clinical applications, and results. *Int Ophthalmol Clin* 1979;**19**:127–89.
13 **Siatkowski RM**, Capo H, Byrne SF, *et al*. Clinical and echographic findings in idiopathic orbital myositis. *Am J Ophthalmol* 1994;**118**:343–50.
14 **Moorman CM**, Elston JS. Acute orbital myositis. *Eye* 1995;**9**:96–101.
15 **Weinstein GS**, Dresner SC, Slamovits TL, *et al*. Acute and subacute orbital myositis. *Am J Ophthalmol* 1983;**96**:209–17.
16 **Patrinely JR**, Osborn AG, Anderson RL, *et al*. Computed tomographic features of nonthyroid extraocular muscle enlargement. *Ophthalmology* 1989;**96**:1038–47.
17 **Mannor GE**, Rose GE, Moseley IF, *et al*. Outcome of orbital myositis. Clinical features associated with recurrence. *Ophthalmology* 1997;**104**:409–13; discussion, 14.
18 **Mombaerts I**, Koornneef L. Current status in the treatment of orbital myositis. *Ophthalmology* 1997;**104**:402–8.
19 **Trokel SL**, Hilal SK. Recognition and differential diagnosis of enlarged extraocular muscles in computed tomography. *Am J Ophthalmol* 1979;**87**:503–12.