ACUTE ORBITAL MYOSITIS

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SUMMARY

We examined 9 consecutive cases of unilateral orbital myositis (7 women and 2 men; age range 15–46 years) presenting to Casualty. Only 3 were correctly diagnosed on the first visit. Eight patients exhibited globe retraction in the acute stages and, after treatment with systemic steroids, all made a full recovery. None of the patients had associated systemic disease and all remain well over a 6–12 month follow-up period. Orbital myositis may be a more common condition than previously thought; it can present with a variety of clinical signs, and may be difficult to diagnose in the early stages. The presence of globe retraction on movement of a painful, injected eye is a useful diagnostic sign which indicates inflammation of extra-ocular muscles and is present in the acute stages of the disease; we explain how to elicit globe retraction and suggest a management protocol for these patients.

Orbital myositis can be defined as a non-specific, localised orbital inflammatory process in which any one or more of the extraocular muscles may be involved. The condition may be acute, subacute or recurrent. In the acute form patients complain of a painful eye and may notice diplopia; clinical examination may show a variety of features ranging from minimal conjunctival injection over the involved muscle to extensive conjunctival chemosis, proptosis, eyelid swelling and blepharoptosis. There may be extraocular muscle limitation. The condition may be unilateral or bilateral and the rectus muscles may be involved alone or in association with the oblique muscles.[1,2] Recurrences may involve the same orbit or the contralateral side.[3]

Orbital myositis can mimic a variety of other ocular conditions such as thyroid eye disease, orbital pseudotumour, orbital cellulitis or any condition which causes enlargement of the extraocular muscles.[3,4] Differential diagnosis depends on blood tests and specific radiological investigations.[5,6] In practice the diagnosis of orbital myositis is made on the clinical picture and confirmed by CT scan. Specific MRI appearances have been described enabling a precise diagnosis to be made and the differentiation of myositis from orbital neoplasia (e.g. lymphoma) and pseudotumours.[7]

Tissue biopsy is rarely indicated in 'typical' cases. Open muscle biopsy or fine needle aspiration biopsies have shown non-granulomatous inflammatory changes with marked lymphocytic infiltration.[8,9]

The aetiology of orbital myositis is not clear although it has been reported in association with many conditions, including respiratory tract infection, myocarditis, Lyme disease, herpes zoster and Whipple's disease.[10–14] Weinstein *et al.*[9] reported 12 patients with orbital myositis 4 of whom had histories of ocular or systemic autoimmune disease.

Treatment of acute orbital myositis with high-dose systemic corticosteroids usually results in prompt clinical improvement; non-steroidal anti-inflammatory agents such as indomethacin are also effective.[15] Subacute or chronic cases which prove refractory to this regime have been shown to respond to second-line anti-inflammatory agents such as low-dose methotrexate or radiation treatment.[9,16]

We report 9 consecutive cases of acute orbital myositis presenting to the Eye Casualty Department at the Oxford Eye Hospital over a 21 month period. This study shows that the condition may present more commonly to Casualty than previously thought and that with appropriate treatment there is a good long-term prognosis. We present a protocol for the management of these patients.

PATIENTS AND METHODS

The patients presented over a 21 month period to the Oxford Eye Hospital Accident & Emergency Department with the symptoms indicated in Table I, which also shows the initial diagnosis. A provisional clinical diagnosis of orbital myositis was made after re-examination by the authors, when globe retraction was specifically looked for. The sign was elicited by first performing horizontal and vertical eye movements with the patient facing the examiner

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Table I. Presenting symptoms and initial diagnosis in patients with orbital myositis (n=9)

Patient no., age (yr) and sex	PMH	Presentation				Duration of symptoms (days)	Initial diagnosis
		Redness	Pain	Diplopia	Headache		
1. 18 F	No	++	++	+	−	5	Conjunctivitis
		+	+	−	−	2	
		−	+	−	−	4	
2. 30 F	No	+	+	+	−	14	Episcleritis
3. 46 F	Crohn's disease	+	−	+	+	21	Episcleritis
4. 25 M	No	++	+	+	−	30	Orbital myositis
5. 36 M	No	+	+	+	−	7	Allergy; orbital cellulitis
6. 36 F	No	−	+	+	+	5	Orbital myositis
7. 15 F	Migraine	+	+	−	−	1	Episcleritis
8. 26 F	No	−	++	−	−	3	Orbital myositis
9. 31 F	No	−	++	+	++	35	Thyroid eye disease

PMH, past medical history.

and checking for narrowing of the palpebral fissure, and then repeating eye movements with the patient in profile and looking for retraction of the globe, which usually occurs when the eye is looking in a direction opposite to the direction of action of the involved muscle. Eight patients then had orbital CT scanning to confirm the diagnosis; 1 patient had an orbital ultrasound scan. Systemic investigations were undertaken as shown in Results. Treatment was initiated as indicated in Table II.

RESULTS

The mean age of the patients at presentation was 29 years (range 15–46 years). Eight of the patients had a history of acute onset, persistent ocular discomfort beginning 1–35 days earlier.

All patients were in good general health with no relevant family history. Ocular examination revealed normal visual acuity, pupil responses, colour vision testing, intraocular pressure and dilated fundal appearance in all patients. There was no sign of intraocular inflammation. Eight patients had globe retraction with narrowing of the palpebral fissure in the acute stages (see Fig. 1).

The patients with presumed conjunctivitis and episcleritis were all reviewed within 2–4 days and on subsequent examination were found to have an orbital condition. The patient with orbital cellulitis was admitted with this diagnosis and treated as such until a CT scan revealed the typical features of myositis. Five of the patients were eventually admitted to hospital once the diagnosis of orbital myositis had been made.

All the patients underwent full physical examination including neurological examination and were found to be healthy; blood tests for full blood count and blood film, urea, electrolytes, blood sugar, liver function tests, erythrocyte sedimentation rate, C-reactive protein, thyroid function tests, VDRL and an auto-antibody screen showed no abnormality. None of the patients has developed systemic disease over a 6–24 month follow-up period.

Hess charts were performed on 7 patients; the 2 who did not have orthoptic follow-up had no diplopia despite slight restriction in adduction of the affected eye.

A CT scan was performed on 8 patients as a diagnostic procedure and in all cases showed swelling of the insertion and the belly of the extraocular muscle involved (Fig. 2) which was consistent with orbital myositis. Dysthroid eye disease has a similar appearance on CT scan but the muscle insertion is usually spared and several muscles in both orbits normally show some change. One patient had an ultrasound scan (orbital B-scan) which showed enlargement of the belly of the involved muscle with reduced reflectivity on the A scan (Fig. 3). These are typical features of orbital myositis, in contrast to thyroid eye disease which would show normal or increased reflectivity on scanning.

Table II. Clinical features and treatment of patients with orbital myositis (n=9)

Patient no.	Diagnostic delay (days)	Proptosis	Globe retraction	Muscle affected	Steroid treatment	Recurrent attacks
1.	5	No	Yes	Left MR, right LR, left LR	20 mg b.d.	2
2.	3	No	Yes	Left LR	20 mg b.d.	0
3.	16	Yes	Yes	Right LR	30 mg b.d.	0
4.	–	No	Yes	Left Mr	None	0
5.	4	Yes	Yes	Left LR	40 mg b.d.	0
6.	–	No	Yes	Right LR	20 mg b.d.	1
7.	11	No	Yes	Left LR	None	0
8.	–	No	Yes	Right LR	20 mg o.d.	0
9.	8	No	No	Left SO	30 mg o.d.	0

MR, medial rectus; LR, lateral rectus; SO, superior oblique.

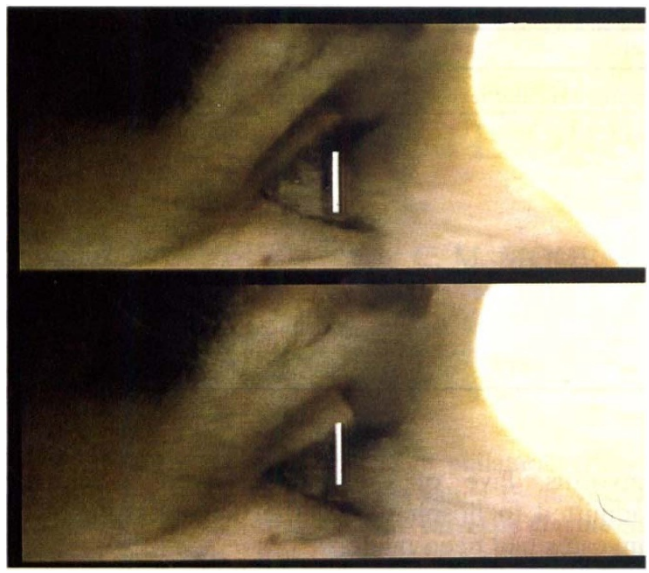

Fig. 1. *Patient 3. Lateral view of the right orbit with the patient in primary gaze (above) and in adduction (below) demonstrating narrowing of the palpebral fissure. Globe retraction was present, but due to the dynamic nature of this sign it is difficult to demonstrate in print.*

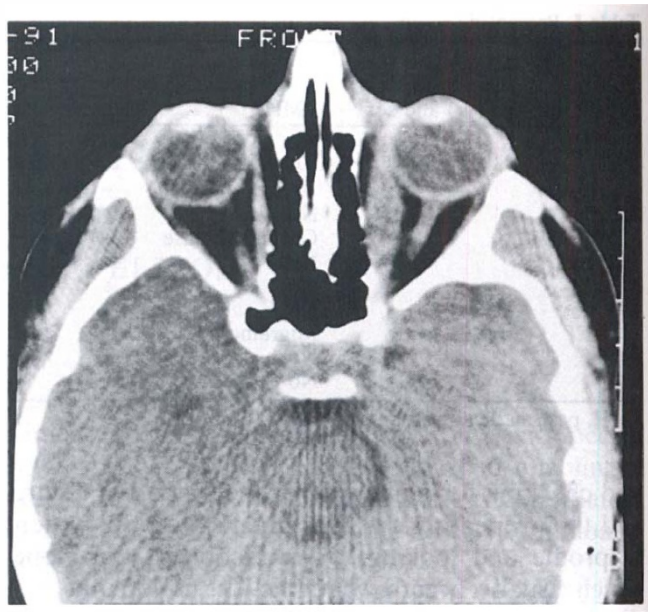

Fig. 2. *Patient 1. CT scan of the orbits showing marked enlargement of the left medical rectus from the insertion to the orbital apex.*

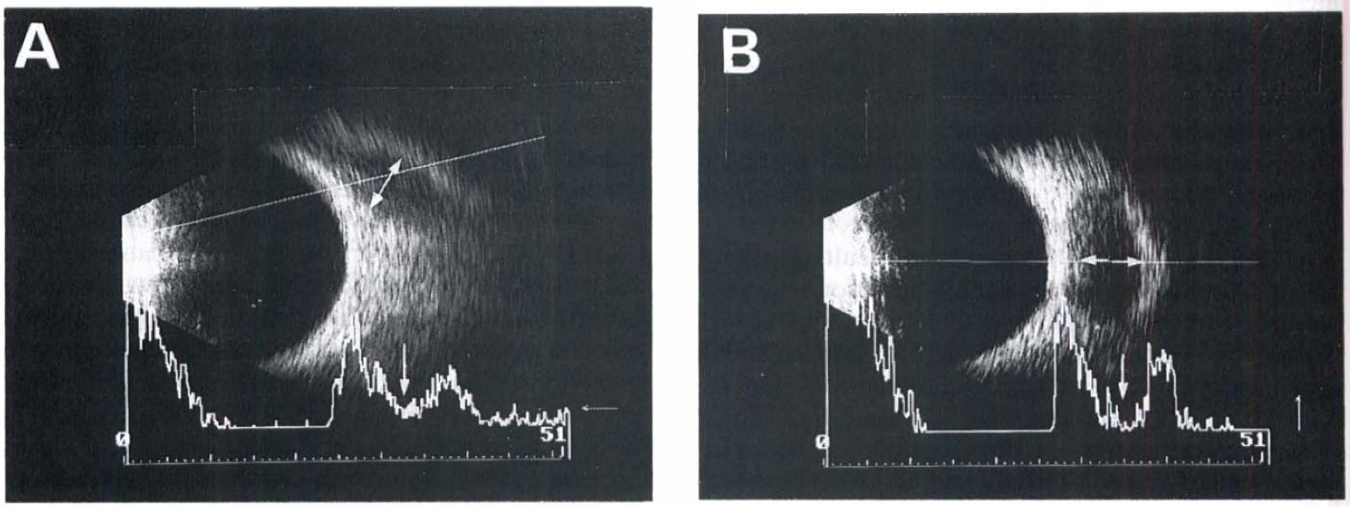

Fig. 3. *Patient 8. A- and B-mode echography of the right lateral rectus. The A scans show marked reduction of internal reflectivity (single arrows). The B-mode echograph shows enlargement of the muscle belly in the horizontal (A) and the vertical (B) plane (double arrows).*

The CT scan can be difficult to interpret in cases involving the oblique muscles and patient 9 was initially thought to have enlargement of the left medial rectus; careful examination of the patient's eye movements and an ultrasound scan showed that the superior oblique alone was affected.[2] It was interesting that despite clinically normal-looking eyes patients 6 and 8 both demonstrated globe retraction and had diagnostic CT and ultrasound scans.

Seven patients had full eye movements and resolution of proptosis at the final outpatient visit; 1 patient had some restriction of eye movements in abduction but was not troubled by this.

DISCUSSION

All our patients had acute orbital myositis confirmed by either CT or ultrasound scan appearances of the involved muscles and the response to treatment. Neither open nor fine needle biopsies were considered necessary. However, only 3 of the 9 patients were correctly diagnosed at the first casualty visit due to a combination of low awareness of the condition and the non-specific clinical signs present. In 1 case no treatment was instituted and the condition resolved completely spontaneously. This suggests that orbital myositis may escape diagnosis and that it may be more common than supposed. When

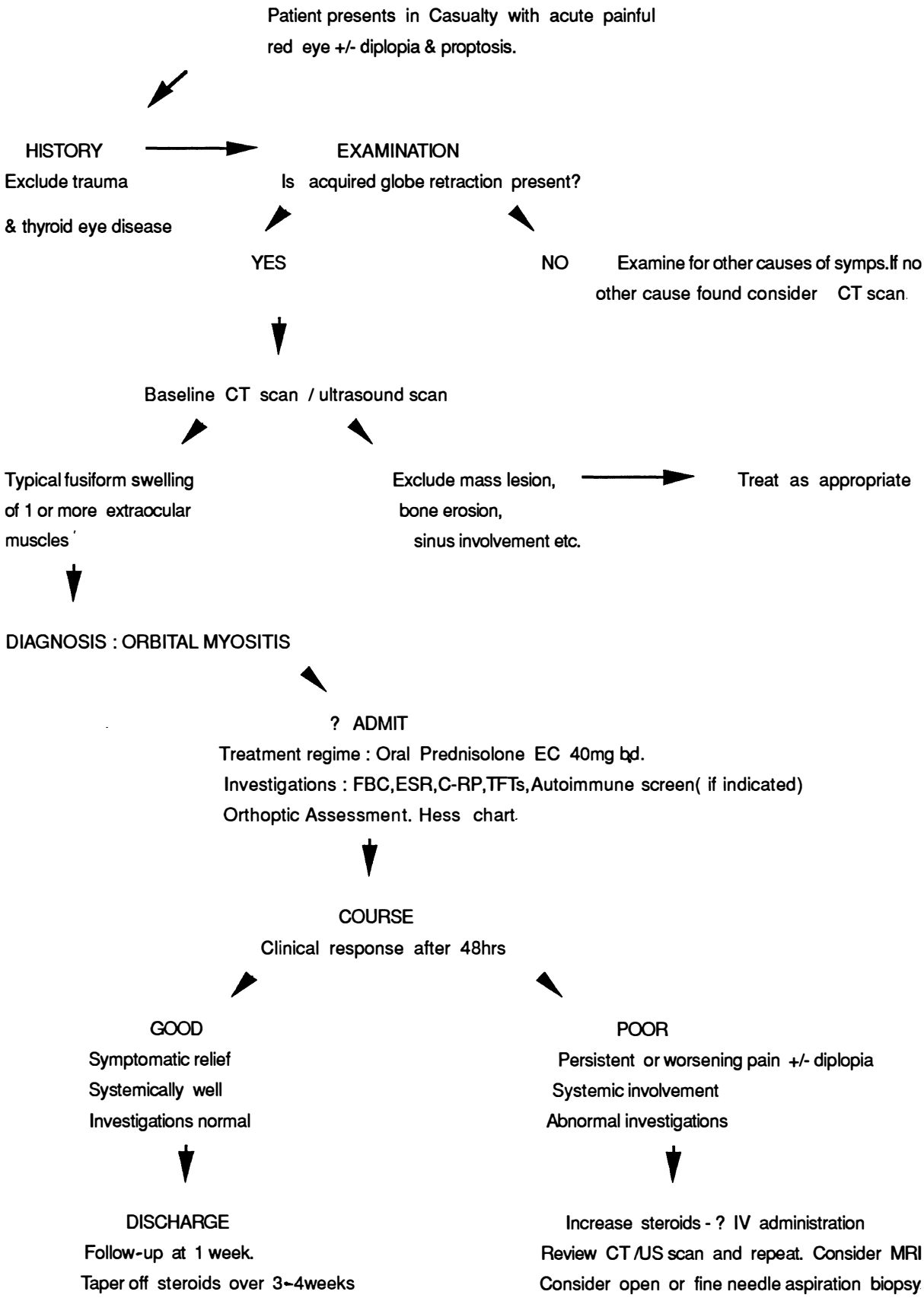

Fig. 4. *Management protocol for patients with acute orbital myositis.*

examined at follow-up, all 9 patients were found to have limited movement of the involved eye and 8 had globe retraction on attempted movement in the opposite direction. This sign localises the pathology to the orbit as opposed to the more anterior processes, such as episcleritis, diagnosed in these patients (Table I).

Acquired globe retraction with narrowing of the palpebral fissure implies infiltration of one of the extraocular muscles by inflammatory myositis, neoplasm or dysthyroid eye disease.[17] It also occurs in the general fibrosis syndrome and blow-out fractures. Congenital globe retraction is a characteristic of Duane's syndrome, in which there is electrophysiological evidence that it is due to co-contraction of the horizontal rectus muscles. Co-contraction may also contribute to globe retraction when it occurs in dysthyroid eye disease. Work done on patients with Graves' ophthalmopathy has shown increased tension and reduced elasticity of involved extraocular muscles with length tension measurements indicating that this is due primarily to active muscle contraction.[18] Attempted movement of the globe in the opposite direction therefore evokes active contraction in the involved muscle, possibly because muscle spindle sensitivity characteristics are altered, generating inappropriate motor neurone activity. Alternatively, globe retraction may be due to muscle stiffness or failure of relaxation. This is the explanation for the phenomenon in orbital blow-out fracture and neoplastic infiltration of extraocular muscles and probably a contributory factor in Graves' ophthalmology. Both co-contraction ('muscle spasm') and muscle entrapment or infiltration will produce a positive traction test. In the case of orbital myositis, it is likely that both mechanisms are involved in producing globe retraction.[19]

Previous studies have reported an association between acute orbital myositis and systemic or generalised autoimmune disease,[9,10,12,14] but in those cases orbital myositis was not the presenting feature of the condition, which had usually been diagnosed for some time. The patients in this study were not found to have any systemic associations at presentation and have continued to remain in good health on follow-up. There does not, therefore, appear to be any indication for extensive systemic investigation of healthy patients whose only findings are compatible with a diagnosis of acute orbital myositis.

Once commenced on systemic corticosteroids all our patients showed some improvement after 48 hours with relief of symptoms such as pain, headache and diplopia and objective improvement in the clinical appearance of the eye. A short course of oral steroids (1–2 weeks) was gradually tapered off over 3–4 weeks. Specific treatment protocols for generalised orbital inflammatory disease, possibly associated with systemic disease, have been suggested, based on very high-dose steroids (either oral, intravenous or intralesional) followed by radiotherapy and/or cytotoxic drugs in resistant cases.[20]

We present a management protocol for patients presenting to Casualty with acute symptoms (Fig. 4). Previous trauma and systemic disease must be excluded in patients with an acute red eye and a routine examination, including eye movements, will reveal the underlying cause in the majority of cases. If globe retraction is elicited during examination a CT scan should be organised from Casualty; if the diagnostic features of orbital myositis are present the patient should be commenced on oral prednisolone 40 mg b.d. and admitted to hospital if necessary. Further investigations are only indicated if the history or general examination suggest underlying pathology or if the CT scan appearances are not typical. Although a CT scan was our investigation of choice in the majority of patients, ultrasonography also demonstrates characteristic features of orbital myositis such as prominent swelling of the insertion and reduced reflectivity of the belly of the muscle, and is extremely useful in differentiating orbital myositis from other causes of enlarged extraocular muscles.[21]

The majority of patients have diplopia and demonstrable muscle restriction so baseline orthoptic assessment with serial Hess charts is useful to monitor the course of the disease. If the patient does not improve clinically on adequate treatment the steroids can be increased as necessary and intravenous administration considered; supplemental radiation therapy may be indicated in refractory cases. We have found that with prompt diagnosis and adequate treatment the clinical prognosis is very good.

Key words: Globe retraction, Management protocol, Orbital myositis.

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