Orbital Myositis: Evaluating Five New Cases Regarding Clinical and Radiological Features

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ABSTRACT

Orbital myositis (OM) is an inflammatory disorder of the extraocular muscles. The signs and symptoms of OM are periorbital pain, eyelid swelling and redness, restricted ocular motility, and strabismus. There are at least two major forms, described by Benedikt GH Schoser; a limited oligosymptomatic ocular myositis (LOOM), which is associated with conjunctival injection only, and severe exophthalmic ocular myositis (SEOM), which presents with additional ptosis, chemosis, and proptosis. We report the clinical and radiological features of five patients with OM who were recently followed in our clinic. Three patients, one man and two women, were placed in the LOOM group, and the other two patients, both women, were in the SEOM group. In both groups, the initial complaints were pain worsening with eye movements and

double vision, with only one patient in the SEOM group having pain worsening secondary to Crohn's disease. The most affected muscles were the medial and lateral recti. All the patients were treated with corticosteroids, resulting in rapid improvement. Only one patient in the SEOM group experienced a relapse. Orbital magnetic resonance imaging of all the patients revealed enlargement and contrast enhancement of the involved muscles. Although clinical and radiological features are quite consistent, delayed diagnosis in some patients demonstrates the importance of the awareness of OM.

Keywords: Orbital myositis, diplopia, extraocular muscles

INTRODUCTION

Orbital myositis (OM) is an autoimmune disorder that is characterized by the inflammation of extraocular muscles. It presents with a limited clinical course and is considered to be a subtype of orbital inflammatory syndrome (1,2). This usually idiopathic disease is more common in middle-aged women. In adults, the disease may appear following viral upper respiratory tract infections or during the course of inflammatory intestinal diseases. It is very rare in the pediatric age group, although infants may be affected (3). Usually, the course is acute or subacute. The principal features are periorbital pain and edema, diplopia, chemosis, ptosis, and gaze paresis (4,5). The disease may be assessed in two subgroups. When only conjunctival hyperemia is associated with extraocular muscle involvement, it is referred to as limited oligosymptomatic ocular myositis (LOOM). If ptosis, proptosis, and chemosis are also additionally found, it is severe exophthalmic ocular myositis (SEOM) (6). Among a series of disorders that are characterized by the enlargement of intraorbital structures, mainly extraocular muscles, thyroid orbitopathy, lymphoproliferative diseases, metastatic conditions, parasitic infections, orbital cellulitis, vasculitis, and arteriovenous malformations and OM can be listed (7). Systemically applied corticosteroid treatment is the most effective management for patients with acute OM; immunosuppressant treatment is the second option (8).

CASES

We assessed the clinical and radiological features of five patients with OM, evaluated their response to treatment, and discussed our findings in light of the relevant literature.

Five consecutive patients with OM who were monitored as inpatients between February 2012 and July 2014 were retrospectively assessed for their demographic features and clinical and radiological findings. The results are presented in tables. Informed consent was obtained from all the patients.

Case 1

A 37-year-old female patient was admitted with pain around the right eye, double vision, and edema in the eyelid. These complaints had appeared 1 month ago and had progressively deteriorated. Moreover, she described similar, separate attacks occurring 7 and 4 years previously, including several brief painful periods over the last 4 years. She was evaluated in detail and was diagnosed and treated for migraine and cluster headache without benefit.

In the neurological examination, chemosis, proptosis, and periorbital edema were noticed. The right eye revealed a slight inward deviation (Figure 1a). The gazes in all directions were limited. An attempt to look in any direction elicited pain. Magnetic resonance imaging (MRI) revealed enlargement of all extraocular muscles, more marked in the right medial and inferior recti (Figure 1b). An OM diagnosis was confirmed, and intravenous methyl prednisolone/1 gram daily was initiated. After a 5-day course, the treatment was switched to oral 1 mg/kg prednisolone daily.

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After 3 months, prednisolone was discontinued in a tapering regimen. In this period, the patient's complaints decreased and her gaze palsies remarkably improved. At the end of 1 year, she visited our clinic again with chemosis, proptosis, and painful external ophthalmoplegia in her right eye. However, no benefit was observed with corticosteroid use on this occasion, and 1 mg/kg/day azathioprine was initiated as an immunosuppressant treatment. After a 3-month treatment, the clinical picture significantly improved.

Case 2

A 48-year-old female patient was admitted with double vision, pain, and redness around her left eye. Her complaints had begun 3 weeks ago and had deteriorated with time. In the neurological examination, limited abduction of the left eye was the sole abnormal finding. MRI revealed thickening of the lateral rectus muscle together with contrast enhancement (Figure 2). Oral 1 mg/kg prednisolone daily was initiated. On the fifth day of treatment, the clinical symptoms completely resolved.

Case 3

A 23-year-old female complained with pain and redness around her left eye and double vision. Her complaints had been ongoing for the past 3 days. The neurological examination revealed chemosis, proptosis, and periorbital edema on the left side (Figure 3a). Abduction of the left eye only was limited. An orbital MRI revealed the enlargement of the left medial rectus muscle that was associated with gadolinium enhancement (Figure 3b). The patient had been diagnosed with Crohn's disease 2 years ago and had experienced an attack 2 weeks ago. We initiated treatment with oral 1 mg/kg prednisolone daily with a diagnosis of secondary OM. Her complaints and gaze paralysis completely resolved on the third day of treatment. After consulting with a gastroenterologist, we decided to continue the prednisolone treatment with the smallest dose possible.

Case 4

A 36-year-old male was examined for pain around his left eye. The pain had begun 5 days ago and had gradually increased to a level where analgesic medications were ineffective. In the neurological examination, the right eye could not completely abduct (Figure 4a). We observed a thickened and contrast-enhanced right medial rectus muscle in the orbital MRI (Figure 4b). A diagnosis of OM was made, and treatment with oral prednisolone was initiated. The pain and gaze paresis completely disappeared in 5 days.

Case 5

A 62-year-old female was admitted with diplopia and pain around her eyes. Similar complaints had been intermittently appearing for 20 years, which responded to analgesics or spontaneously resolved. During the first

examination, the patient's eye movements could not be completely examined because of severe pain. MRI revealed the enlargement and contrast enhancement of both medial recti muscles (Figure 5b). High-dose (1 gram daily) intravenous methyl prednisolone was initiated with a presumed diagnosis of OM. On the second day, the pain was relieved; thus, a full examination could be completed. The right eye was laterally deviated, and its movements were limited in all planes, more remarkably horizontally (Figure 5a). The treatment was maintained for 7 days and then gradually tapered. In the follow-up visit after 1 month, the pain had completely passed, and the patient's gaze paresis was resolved.

The patients (cases 1 and 3) who had chemosis, proptosis, periorbital edema, and extraocular muscle involvement were included in the SEOM group, and the others patients who had only conjunctival hyperemia and extraocular muscle involvement were included in the LOOM group.

The mean ages of the patients were 40 (23–62), 49 (36–62), and 30 (23–37) years for the whole group, LOOM group, and SEOM group, respectively. Two patients in the LOOM group were women and one was a man, while both patients in the SEOM group were women. In both groups, the initial complaints were pain worsening with eye movements and double vision (Table 1). While one patient in the SEOM group had pain worsening secondary to Crohn's disease, all others were considered idiopathic. The clinical features of patients with OM as a whole and in the LOOM and SEOM groups are separately provided in Table 2, and the distribution of the extraocular muscle involvement is provided in Table 3. All the patients were treated with corticosteroids, resulting in rapid

Figure 3. a, b. (a) Limited abduction of the left eye with chemosis, proptosis and periorbital edema, (b) Orbital MRI: Enlargement of the left medial rectus muscle, with contrast enhancement

Figure 4. a, b. (a) Limited abduction of the right eye. (b) Orbital MRI: Enlargement and contrast enhancement of the right medial rectus muscle

Figure 5. a, b. (a) Laterally deviated right eye together with limitation of both eye movements in all planes. (b) Orbital MRI: Enlargement and contrast enhancement of both medial recti muscles

improvement. However, one patient in the SEOM group experienced a quick recurrence, which necessitated immunosuppressive treatment with azathioprine (Table 2).

DISCUSSION

The most common diseases of the orbit are thyroid opthalmopathy, lymphoproliferative disorders, and OM (7). We here present five consecutive cases that we consider worthy of reporting with their differentiating features. The majority of patients were females of young or middle age (four women to one man). Periorbital pain worsening with eye movements was the initial complaint in all patients. In four patients, double vision was associated with pain (Table 1). The clinical pictures were similar to case reports found in the relevant literature (2,3,4,5,6).

The differential diagnosis of idiopathic OM includes the following: infections (viral infections, orbital cellulitis, orbital abscess, Lyme disease, Herpes zoster, and syphilis), inflammatory reaction (trauma, foreign body, bisphosphonate-related reaction, and postvaccinal reaction), Tolosa–Hunt syndrome, thyroid ophthalmopathy, vasculitis (Wegener's granulomatosis, polyarteritis nodosa, rheumatoid arteritis, giant cell arteritis, and Kawasaki disease), systemic lupus erythematosus, sarcoidosis, inflammatory bowel disease, neoplasm, arteriovenous fistulas, and malformations (3). Because idiopathic OM is a diagnosis of exclusion after physical examination and history taking, all patients underwent imaging and laboratory tests, including complete blood count, hepatic and renal function tests, erythrocyte sedimentation rate, C-reactive protein level, thyroid function tests, various antibodies (such as antineutrophil cytoplasmic, anticardiolipin, antiphospholipid, anti-DNA, antinuclear, anti-Sm, and anti-SCL-70), 175

Table 1. Demographic characteristics and symptoms

Case	Age (years)	Sex	Systemic disorders	Erythema-Edema	Pain	Diplopia	Duration of symptoms
1	37	Female	-	+	+	+	30 days
2	48	Female	-	-	+	+	20 days
3	23	Female	Crohn's disease	+	+	+	3 days
4	36	Male	-	-	+	-	5 days
5	62	Female	-	-	+	+	10 days

Table 2. Clinical features

Results	LOOM	SEOM	LOOM+SEOM
Mean age (years)	49 (36–62)	30 (23–37)	40 (23–62)
Sex (Female:Male)	2:1	2:0	4:1
Clinical presentation			
Acute	1/3 (33.3%)	1/2 (50%)	2/5 (40%)
Subacute	1/3 (33.3%)	-	1/5 (20%)
Chronic	1/3 (33.3%)	1/2 (50%)	2/5 (40%)
Idiopathic OM	3/3 (100%)	1/2 (50%)	4/5 (80%)
Secondary OM	-	1/2 (50%)	1/5 (20%)
Response to steroid treatment	3/3 (100%)	2/2 (100%)	5/5 (100%)
Recurrence	-	1/2 (50%)	1/5 (20%)
LOOM: limited oligosymptomatic ocular myositis; SEOM: severe exophthalmic ocular myositis; OM: orbital myositis			

Table 3. Distribution of muscle involvements

Involved muscles	LOOM	SEOM	LOOM+SEOM
Single muscle	2/3 (66.6%)	1/2 (50%)	3/5 (60%)
Multiple muscles	1/3 (33.3%)	1/2 (50%)	2/5 (40%)
Bilateral	1/3 (33.3%)	-	1/5 (20%)
Medial rectus	2/3 (66.6%)	1/2 (50%)	3/5 (60%)
Inferior rectus	-	1/2 (50%)	1/5 (20%)
Lateral rectus	1/3 (33.3%)	2/2 (100%)	3/5 (60%)
Superior rectus	-	1/2 (50%)	1/5 (20%)
LOOM: limited oligosymptomatic ocular myositis; SEOM: severe exophthalmic ocular myositis			

rheumatoid factor, VDRL-RPR, viral markers (such as HSV, HIV, HBV, and HCV), chest X-ray, ECG and echocardiography, thyroid and abdominal ultrasonography, and thorax CT. No abnormal results were found in the whole group.

A special issue worthy of mentioning separately is IgG4-related sclerosing disease (IgG4-RD), a recently described clinical entity. IgG4-RD is characterized by a typical histopathological appearance and an elevated number of IgG4-positive plasma cells within the involved tissue (9). Serum IgG4 elevation is not specific for this disease. The lymph nodes, hepatobiliary tract, orbits, salivary glands, and particularly the pancreas are commonly involved sites. Orbital IgG4-RD is reported to involve not only the lacrimal gland but also other ocular adnexal tissues such as the extraocular mus-

cles, lacrimal sac, orbital adipose tissue, periorbital membranes, and eyelids (10,11). In our study, two patients (Cases 1 and 3) with eyelid and extraocular muscle involvements could have orbital IgG4-RD. However, the patients rejected biopsies; therefore, we could not assess this probability. We continued the corticosteroid medication with beneficial response, as the same treatment modality, immunosuppression, is effective for both diseases (12).

In one of the patients, we considered a secondary evolution due to Crohn's disease. The remaining patients did not describe any systemic disease, which could lead to OM, and systemic investigations did not reveal a specific condition. We believe that these patients may be accepted as having idiopathic OM. The clinical picture began acutely in two patients and subacutely in one. However, it was apparently chronic in two patients, with histories of 7 and 20 years (Table 2).

The patients presented different pictures according to their extraocular muscle involvement. In three patients (60%), a single muscle involvement was detected. In two, multiple extraocular muscles were affected. In only one patient, bilateral involvement was noticed. The muscles most often affected were the medial and lateral recti (Table 3). These findings were also compatible with the literature (6). We did not find oblique muscle affection in any of our patients. Oblique muscle paralysis is reported together with multiple muscle involvements in the relevant literature (13).

A limitation of any of the eye movements points to dysfunction of the affected extraocular muscle. However, as seen in two of our patients (cases 3, 4), although the medial rectus was the affected muscle radiologically, outward gaze was limited. This finding points to the complexity of eye muscle coordination.

Three of our patients were in the LOOM group, and two were in the SEOM group. All of the patients in the LOOM group were idiopathic, and no recurrence was seen. Although there were single, multiple, and bilateral affections of the extraocular muscles in this group, non-involvement of the inferior and superior rectus muscles is noticeable.

One of the patients with a chronic course (Case 1) was in the SEOM group, as was the secondary OM case (Case 3). Furthermore, no patient in the SEOM group had bilateral involvement of the extraocular muscles. The differences cited here, of course, do not have statistical meaning due to the limited number of patients in this series. However, a trend of clustering for some features cannot be denied.

An important finding in our study is the good response to corticosteroid treatment, with nearly complete resolution of features. The beneficial effect of steroids is repeatedly mentioned in the literature (6,7,8).

One patient experienced a relapse (Case 1). As corticosteroids were ineffective, the treatment was switched to an immunosuppressant drug. In

this particular patient, unilateral but multiple muscle involvements were noted. Both unilateral-multiple and bilateral muscle involvements are the most important predictors of recurrence (13,14).

In this series, which consisted of five OM cases, the diversity of the clinical and radiological findings drew our attention. Many features of our patients share common properties with patients in previous reports. As further patients are reported, features unique to our population may appear. A striking matter noticed in this study is the delay of diagnosis in chronic cases, although OM is one of the most common diseases of the orbit. In patients applying with periorbital pain, double vision, and gaze palsies, OM is an important diagnostic option and must always be considered.

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