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Orbital Myositis

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1. Introduction

Orbital myositis is an inflammation of mainly the extraocular muscles. Orbital myositis has a sudden onset, and the clinical course can be acute or chronic. The ocular signs and symptoms of eyes with orbital myositis are periocular pain, eyelid swelling and redness, restricted ocular motility, and strabismus. Computed tomographic (CT) scans show indistinct swelling around one or more extraocular muscles, and fat-suppressed T2-weighted magnetic resonance (MR) images show localized inflammations. The exact etiology of the inflammation has not been determined, however some cases have been reported to be caused by infectious agents while other cases by autoimmunity. Spirochetotic (Lyme disease), viral (herpes zoster virus), and bacterial infections (Group A streptococcal pharyngitis) can cause orbital myositis. Autoimmune-related orbital myositis is associated with relatively specific diseases: giant cell myocarditis, Crohn disease, and linear scleroderma.

Orbital myositis must be differentiated from other diseases that also have extraocular muscular enlargements, e.g., thyroid-associated orbitopathy, lymphoproliferative disorders, metastatic orbital diseases, parasitic infection, systemic anti-neutrophil cytoplasmic antibody-related vasculitis, and inflammatory conditions triggered by medications and foreign bodies.

The first line of treatment of orbital myositis is systemic corticosteroids to which the inflammation responds well especially the acute type. However, there are corticosteroid-resistant chronic types, and immunosuppressive and biological agents can be used in these types.

2. Patients, signs and symptoms, and imaging studies

2.1 Patients

The etiology of orbital myositis has not been completely established. In some cases, the orbital myositis has a specific etiology, which is described in Section 3. Although idiopathic orbital myositis may occur at any age, it is most commonly present in middle-aged patients. Meta-analysis, including the largest published series (Siatkowski et al., 1994), showed that orbital myositis occured most frequently in young to middle-aged patients with a 1 male to 2 female patient ratio (Table 1; Scott & Siatkowski, 1997). However, a relatively large published case series (Lacey et al., 1999) and our study did not reveal a female predominance (Table 1).

It has also been reported that idiopathic orbital myositis can develop in pediatric patients. Pediatric cases of orbital myositis develop secondary to systemic conditions, e.g.,
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streptococcal pharyngitis (Alshaikh et al., 2008; Belanger et al., 2010) and presumed allergic responses (Yan et al., 2006). There are cases of orbital myositis that develop after pregnancy (Hiraga et al., 2008; Mombaerts & Koornneef, 1997) and cases with a familial history (Jacob et al., 2007; Maurer & Zierz, 1999). However, these may be exceptional cases.

Table 1. Epidemiology of patients with idiopathic orbital myositis.

<table>
<thead>
<tr>
<th>References</th>
<th>Patient number</th>
<th>Male/female ratio</th>
<th>Median or mean age (range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scott et al. 1997</td>
<td>190</td>
<td>0.5</td>
<td>37 (3-84) years old</td>
</tr>
<tr>
<td>Lacey et al. 1999</td>
<td>40</td>
<td>0.74</td>
<td>40 (not available) years old</td>
</tr>
<tr>
<td>Our study</td>
<td>43</td>
<td>1.0</td>
<td>47 (23-91) years old</td>
</tr>
</tbody>
</table>

2.2 Signs and symptoms

Idiopathic orbital myositis is characterized by a sudden onset of orbital inflammation, periocular pain, swelling and redness of the eyelids, proptosis, ptosis, and ocular motility restrictions (Figure 1 to 7). It must be differentiated from idiopathic orbital inflammation and orbital cellulitis, because the signs and symptoms are similar. However, idiopathic orbital myositis can sometimes have atypical signs and symptoms, viz., subacute/chronic onset or be a non-inflammatory condition. These cases with atypical signs and symptoms simulate orbital tumors, and they account for 7% of all cases (Rootman, 2003).

Idiopathic orbital myositis must also be differentiated from other diseases that have enlargements of the extraocular muscles as described in Sections 4.3 to 4.10. Idiopathic orbital myositis can affect any extraocular muscles but rarely the superior and inferior oblique muscles (Figure 7; Kau et al., 2006; Stidham et al., 1998). It also rarely affects the levator palpebrae muscle (Figure 1; Almekhlafi & Fletcher. 2008). Ocular motility is typically restricted in the field of action of the affected extraocular muscles, and also in the direction opposite to its field of action (Figure 5; Kubota & Kano. 2007; Lacey et al., 1999; Siatkowski et al., 1994).

Idiopathic orbital myositis can have an acute or chronic/recurrent clinical course. Acute orbital myositis is generally resolved within 2 months after systemic steroid therapy (Figure 5 and 6). However, chronic/recurrent orbital myositis responds poorly to systemic steroid therapy, and the ocular motility is restricted for more than 2 months and often years (Figure 7).

Fig. 1. Idiopathic orbital myositis of left levator palpebrae muscle.

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External photograph of an 82-year-old man showing left ptosis but with normal ocular motility. Fat-suppressed T2-weighted MR image shows a hyperintense signal in the left levator palpebrae muscle (arrowhead). He recovered spontaneously.

Fig. 2. External photographs of patients with idiopathic orbital myositis. Left. A 91-year-old woman showing unilateral chemosis and ptosis, and with ocular motility restrictions. Right. A 26-year-old man with ptosis, ocular motility restrictions, and periorcular pain.

Fig. 3. External photographs of a patient with idiopathic orbital myositis. Left: A 32-year-old woman showing left upper eyelid swelling and redness. Right: Same patient after a bilateral recurrence while being treated with 5 mg maintenance dose of prednisolone and one week after 1000 mg intravenous methylprednisolone for 3 days.

2.3 Imaging studies
CT scans of eyes with idiopathic orbital myositis show indistinct swelling around one or more muscles with no specific pattern of which muscle is enlarged (Figure 2 to 6). On the
other hand, the MR images of acute and chronic/recurrent type of orbital myositis have characteristic patterns. Fat-suppressed T2-weighted MR images showed inflammation of the extraocular muscles rather than the ocular adnexal structures (Ohnishi et al., 1994). Localized inflammation of the extraocular muscles in eyes with idiopathic orbital myositis at the active stage can be seen in Figures 4 to 7. The acute type of idiopathic orbital myositis has localized areas of hyperintense signals around the extraocular muscles and fascicle structures, whereas the chronic/recurrent type shows areas of hyperintense signals in the extraocular muscles (Kubota & Kano, 2007).

Fig. 4. Fat-suppressed T2-weighted MR image and coronal (middle) and axial (right) CT images recorded at the same time in a patient with idiopathic orbital myositis in the active phase. Top: Acute type of idiopathic orbital myositis. Fat-suppressed T2-weighted MR image shows a hyperintense signal in the fascicle structure (arrowhead) around the superior rectus muscle, and indistinct swelling around the superior rectus muscle in the CT image (Top center). An arrow points to the optic nerve for comparison to coronal CT scan at the posterior pole. Bottom: Chronic type of idiopathic orbital myositis. Fat-suppressed T2-weighted MR image shows a hyperintense signal of the superior rectus muscle (arrow) and the adjacent structures (arrowhead). Reproduced with permission from Kubota, T. & Kano, H. (2007) Assessment of inflammation in idiopathic orbital myositis with fat-suppressed T2-weighted magnetic resonance imaging. American Journal of Ophthalmology, Vol.143, No. 4, pp.718-720, ISSN:0002-9394
Fig. 5. Acute type of idiopathic orbital myositis. Left: External photographs of a 58-year-old man showing swelling of the right eyelid, ptosis, and ocular motility restrictions in the field of action and in the direction opposite to the field of action of the affected extraocular muscles. Severe engorgement of the conjunctival and episcleral vessels overlying the right eye. Fat-suppressed T2-weighted MR image at the active phase shows a hyperintense signal around the fascicular structures (arrowhead). Right: Same patient after 1000 mg intravenous methylprednisolone for 3 days (steroid pulse therapy). There is a complete recovery from signs and symptoms. MR images are normal.
Fig. 6. Acute type of atypical idiopathic orbital myositis. Left: External photographs of a 65-year-old woman showing the left blepharoptosis, and also with ocular motility restrictions in the field of action of the affected extraocular muscles. Signs including subacute onset and non-inflammatory conditions indicate that this is atypical idiopathic orbital myositis. Right: CT images show indistinct swelling around the superior rectus muscle of the left eye. Fat-suppressed T2-weighted MR image shows hyperintense signal around the fascicular structures (arrowhead). Following steroid pulse therapy, she completely recovered from the signs and symptoms.

3. Etiological factors

The cause of the orbital myositis is not known in most cases, however several cases with known etiology have been reported. Infectious and autoimmune-related factors that affect the extraocular muscles are described in this section.

3.1 Infectious agents causing orbital myositis

Cases of orbital myositis caused by infectious agents are rare. Spirochetes (Lyme disease), viruses (herpes zoster virus), and bacteria (Group A streptococcal pharyngitis) are microbes that can cause infectious orbital myositis. The signs and symptoms of each infectious agent are similar to those of idiopathic orbital myositis: acute onset, periocular inflammation, periocular pain, conjunctival hyperemia, eyelid swelling, diplopia, and restriction of ocular movements. Imaging studies show that the findings in eyes caused by the different infectious agent are also similar to those of idiopathic orbital myositis. The protocol of treatment depends on the sensitivity profile of each microbe.

Lyme disease is caused by the spirochete *Borrelia burgdorferi* and is mainly reported in the northern and western United States and also in Europe. Ticks transmit the spirochete which
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Fig. 7. Chronic/recurrent type of idiopathic orbital myositis. A 46-year-old woman with left periocular inflammation and ocular motility restrictions was referred to our hospital because her myositis was refractory to high doses of prednisolone with a slow taper. A: Fat-suppressed T2-weighted MR image at the initial examination shows hyperintense signals in the left inferior and lateral rectus, and superior oblique muscles. B: Following steroid pulse therapy, there is a complete recovery of the signs and symptoms. C: and D: and E: She received high dose steroid (prednisolone 60mg) with a slow taper, however the myositis recurred three times during the tapering. Note that more than one muscle including the right superior oblique muscle was affected. F: She developed a strabismus two years after the initial visit and had persistent ocular motility restrictions.

can affect different organ: the skin, nervous system, heart, joints, and muscles (Holmgren & Matteson, 2006; Muller-Felber et al., 1993; Pendse et al., 2006). The infection by Borrelia burgdorferi can be unilateral and more than one extraocular muscle can be infected. Lyme disease also has other ocular manifestations, dacroadenitis (Nieto et al., 2008), periocular inflammation (Carvounis et al., 2004; Holak et al., 2006), and neuro-ophthalmologic manifestations (Lesser et al., 1990; Pendse et al., 2006; Seidenberg et al., 1990). The diagnosis is based on the patients living in or visiting an endemic area, a skin rash, and a positive serologic test for Borrelia burgdorferi. Oral doxycycline is the best treatment, and steroids may also resolve the inflammation.

Herpes zoster is a rare cause of orbital myositis, but several cases have been extracted by a PubMed search (Badilla et al., 2007; Kawasaki & Borruat, 2003; Krasniaanski et al., 2004; Volpe et al., 1991). The signs and symptoms of orbital myositis associated with herpes zoster are similar to those of acute orbital myositis. The characteristic skin rash of herpes zoster may develop after the ocular adnexal inflammatory conditions (Kawasaki & Borruat, 2003). Herpes zoster can be unilateral and can affect more than one extraocular muscle. Acyclovir can improve the ocular manifestations, and cure the disease before progressing to the chronic clinical stage.
Orbital myositis following streptococcal pharyngitis is also a rare condition. Several cases have been reported based on a PubMed search (Alshaikh et al., 2008; Belanger et al., 2010; Culligan et al., 2005; Purcell et al., 1981). The highest incidence of this type of orbital myositis is in infants and young adults. In general, the orbital myositis occurs two to six weeks after the development of streptococcal pharyngitis. The pathogenesis may have an immunocompromise factor rather than streptococcal A. Imaging studies show unilateral with single or multiple extraocular muscular enlargements. The orbital myositis following streptococcal pharyngitis responds to oral corticosteroids, and can be cured without progressing to the chronic stage.

3.2 Orbital myositis associated with autoimmunity

Cases of orbital myositis associated with autoimmunity are associated with relatively specific autoimmune diseases; giant cell myocarditis, Crohn disease, systemic lupus erythematosus, rheumatoid arthritis, and linear scleroderma. Idiopathic giant-cell myocarditis is a rare and fatal disorder. Relatively young adults (mean age 43 years-old) are affected, and they usually die of heart failure and ventricular arrhythmia (Cooper et al., 1997). Nineteen percent of patients are associated with autoimmune disorders: Crohn disease, ulcerative colitis, and orbital myositis (Cooper et al., 1997). Five cases of orbital myositis associated with giant cell myocarditis have been published based on a PubMed search (Kattah et al., 1990; Klein et al., 1989; Leib et al., 1994; Lind-Ayres et al., 2009; Stevens et al., 1996). The patients were 14-to 65-years-old and all were women. Their signs and symptoms are similar to those of patients with idiopathic orbital myositis, and patients present with peri-orbital pain, proptosis, ptosis, ocular motility restrictions, and swelling of the extraocular muscles including their tendons. Steroid treatments improve their signs and symptoms, but patients can develop cardiogenic episodes usually within a couple of months of onset of orbital myositis. Therefore, physicians should consider the possibility of idiopathic giant-cell myocarditis especially when a young woman is diagnosed with idiopathic orbital myositis.

Inflammatory bowel diseases, e.g., Crohn disease and ulcerative colitis, have ocular manifestations in 4% to 12% of the cases (Ghanchi et al., 2003). The signs and symptoms include scleritis, uveitis, neuro-ophthalmic, corneal and retinal complications, and orbital pseudotumor. Orbital inflammation is believed to be a true complication of inflammatory bowel disease (Ghanchi et al., 2003). Orbital inflammation rapidly responds to systemic steroids but a reduction of the steroid dose may lead to recurrences.

Scleroderma is a chronic autoimmune disease previously called the CREST syndrome and it has cutaneous manifestations that affect the arms and face. It is characterized by fibrosis, vascular alternations, and autoantibodies. The ocular manifestations involve the extraocular muscles which can appear atrophic (Suttrop-Schulten & Koornneef, 1990) or enlarged (Ramboer et al., 1997).

Several cases of orbital myositis associated systemic lupus erythematosus (Grimson et al., 1983; Serop et al., 1994) and rheumatoid arthritis have been published (Nabili et al., 2002; Panfilio et al., 2000).

4. Differential diagnosis

Orbital myositis is characterized by periocular and/or orbital inflammations and extraocular muscle enlargements. Various diseases must be differentiated from orbital
myositis. First, periocular and/or orbital inflammations of idiopathic orbital inflammation and orbital cellulitis are similar to those of orbital myositis. Second, signs and symptoms of thyroid-associated orbitopathy have also similar to those of orbital myositis and it has high incidence among the orbital diseases. Finally, primary and secondary carcinoma, lymphoproliferative lesions, parasite infection, anti-neutrophil cytoplasmic antibody-mediated systemic vasculitis, some kind of drugs and foreign bodies can lead to extraocular muscle enlargements.

4.1 Idiopathic orbital inflammation
Idiopathic orbital inflammation is caused by unknown etiology, and inflammatory conditions are specific to the ocular adnexa: a lacrimal gland, an eyeball, extraocular muscles, and an optic nerve (Figure 8). This may be due to a number of different organ-specific immunologic disorders of more specific etiologies yet to be defined (Rootman, 2003). Imaging studies can differentiate each type. However, periocular type and idiopathic orbital myositis arising from superior or inferior extraocular muscles may often be difficult to differentiate between them.

Fig. 8. CT images of each type of idiopathic orbital inflammation. A: orbital myositis, B: periopotic type, C: periocular type, D: apical type, E: lacrimal type, F: diffuse type

4.2 Orbital cellulitis
Orbital cellulitis is the inflammation caused by bacterial infections. It may be difficult to differentiate eye with orbital cellulitis from orbital myositis especially at early stage. Therefore, the diagnosis orbital myositis is often made following initial therapy for potential infectious etiology (Costa et al., 2009). In general, idiopathic orbital myositis characterizes a sudden onset. In addition, signs and symptoms reach their peak intensity at the initial onset. In contrast, orbital cellulitis characterizes an acute onset, but progressively develops signs and symptoms. Even though signs and symptoms resemble that of orbital myositis at active phase of orbital cellulitis (Figure 9), imaging studies may help a differential diagnosis between them. CT scans of idiopathic orbital myositis reveal extraocular muscle enlargements, whereas those of orbital cellulitis reveal fuzzy diffuse pattern in the orbit (Figure 9). However orbital cellulitis often progressive to massive lesions (Rootman, 2003). MR images of idiopathic orbital myositis reveal localized inflammations specific to affected extraocular muscles, whereas, those of orbital cellulitis reveal same signals in the vitreous body due to abscess formations that extend diffusely to the orbit (Figure 9).
4.3 Thyroid-associated orbitopathy

Thyroid-associated orbitopathy is the most specific disease that shows extraocular muscle enlargements. The differential diagnosis thyroid-associated orbitopathy and orbital myositis are shown in Table 2. Patients with thyroid-associated orbitopathy typically lack pain, have eyelid retraction with lagophthalmos, thyroid associated autoantibodies, and sparing of the extraocular muscle tendons on imaging (Figure 10). However, the signs and symptoms may overlap those of orbital myositis. For example, idiopathic orbital myositis without tendon involvement has been reported (Patrinely et al., 1989).

<table>
<thead>
<tr>
<th></th>
<th>Idiopathic orbital myositis</th>
<th>Thyroid-associated orbitopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Onset</strong></td>
<td>Sudden, acute</td>
<td>Subacute, chronic</td>
</tr>
<tr>
<td><strong>Periocular inflammatory signs</strong></td>
<td>Frequent</td>
<td>Infrequent</td>
</tr>
<tr>
<td><strong>Bilaterality</strong></td>
<td>Infrequent</td>
<td>Frequent</td>
</tr>
<tr>
<td><strong>Eyelid</strong></td>
<td>Limitation in the field of action and often in the direction opposite to the field of action of the affected extraocular muscles</td>
<td>Limitation in the direction opposite to the field of action of the affected extraocular muscles</td>
</tr>
<tr>
<td><strong>Extraocular movements</strong></td>
<td>opposite to the field of action of the affected extraocular muscles</td>
<td>Distinct swelling and infrequent tendon involvement</td>
</tr>
<tr>
<td><strong>CT image</strong></td>
<td>Indistinct swelling and frequent tendon involvement</td>
<td>A hyperintense signal around fascicle structures in acute type or a hyperintense signal in the muscle in chronic type</td>
</tr>
<tr>
<td><strong>Fat-suppressed T2-weighted MR image</strong></td>
<td>A hyperintense signal in the muscle</td>
<td>A hyperintense signal in the muscle</td>
</tr>
<tr>
<td><strong>Thyroid-associated autoantibodies</strong></td>
<td>Negative</td>
<td>Frequently positive</td>
</tr>
<tr>
<td><strong>Response to steroids</strong></td>
<td>Dramatic with complete resolution in acute type and incomplete or recurrent in chronic type</td>
<td>Incomplete and slow</td>
</tr>
</tbody>
</table>

Table 2. Differential diagnosis of idiopathic orbital myositis versus thyroid-associated orbitopathy.
4.4 Primary and metastatic tumors

In a large cohort case series of orbital tumors and simulating lesions in the United States, Canada, and Japan, ocular adnexal lymphoid tumors accounted for approximately 10% to 18% of all cases (Garrity et al., 2007; Rootman, 2003; Shields et al., 2004; Shikishima, et al., 2006). Among malignant lymphomas, marginal zone B cell lymphomas made up the majority of the lymphomas arising from the ocular adnexa (Ferry et al., 2008). Among benign lymphoproliferative disorders, ocular adnexal IgG4-related related lymphoplasmacytic infiltrative disorder appeared to be a separate clinical entity that has unique clinical characteristics (Kubota et al., 2010). The signs and symptoms of them are frequently similar to those of atypical idiopathic orbital myositis (Figure 11 and 12). When patients have their signs and symptoms, physicians should consider either atypical idiopathic orbital myositis or ocular adnexal lymphoproliferative disorders. They should also consider an incisional biopsy.

In the cohort case series studies, secondary or metastatic tumors account for approximately 20% to 40% of all cases (Garrity et al., 2007; Rootman, 2003; Shields et al., 2004; Shikishima, et al., 2006). Any malignant tumors in the body have a potential to metastasize to the extraocular muscles, and malignant tumor from the lung, breast, and thyroid were found to predominate (Shikishima et al., 2006).

In imaging studies, the extraocular muscles in eyes with idiopathic orbital myositis appear indistinct and enlarged. On the other hand, metastatic tumors to the extraocular muscle appear sharply defined with irregular extraocular muscle enlargement (Figure 13). However, the images also resemble those of eyes with orbital myositis (Capone & Slamovis, 1990; Devine & Anderson, 1982; Slagle et al., 2009).

Fig. 11. Primary marginal zone B-cell lymphoma arising from left lateral rectus muscle. A 62-year-old man had periocular pain and ocular motility restrictions at initial visit. After steroid pulse therapy, he recovered from ocular symptoms but conjunctival mass lesions and extraocular muscle enlargements remained.
Fig. 12. Secondary follicular lymphoma. CT image of the orbit of a 57-year-old man with a history of follicular lymphoma had extraocular enlargements. Note that ocular motility is within normal limits despite of extraocular enlargements.

Fig. 13. Secondary carcinomas metastatic to extraocular muscles. Cases of breast carcinoma (left) and gastric carcinoma (Right) that metastasized to the orbit. Note that definitive and irregular enlargements of the extraocular muscle (arrowheads).

4.5 Parasitic infections
Two species of parasitic worms can infect the extraocular muscles; orbital sparganosis is reported mainly in eastern Asia and orbital cysticercosis mainly in India. Orbital sparganosis is caused by *Spirometra erinaceieuropaei* and can be acquired by drinking water containing copepods infected with the larval stage of the parasite. The orbit is a favorable site (Wiwanitkit, 2005; Yoon et al., 2004), and sparganosis can infect the extraocular muscles. (Figure 14; Kubota & Itoh, 2007). It is difficult to distinguish orbital myositis associated with sparganosis from idiopathic orbital myositis. A presumptive diagnosis of sparganosis can be made by finding a painful migratory subcutaneous nodule (Markell et al., 1999). An accurate diagnosis is made following the surgical removal and identification of the worm.

Orbital cysticercosis is caused by a parasitic *Cysticercus cellulosae* infection which can infect the extraocular muscles. The host for *C. cellulosae* is the pig, and patients usually acquire the infection by eating undercooked pork. Imaging studies of orbital cysticercosis are
characteristic, and the findings can differentiate of orbital cysticercosis from idiopathic orbital myositis (Angotti-Neto et al., 2007; Pushker et al., 2002; Rath et al., 2010)


4.6 Anti-neutrophil cytoplasmic antibody (ANCA)-related vasculitis
Patients with systemic ANCA-related vasculitis, e.g., Churg-Strauss syndrome and Wegener granulomatosis, often also have pseudotumors in the ocular adnexae. The pseudotumors may resemble orbital myositis (Figure 15; Fujii et al., 2010; Takanashi et al., 2001). The ocular symptoms are often intolerable periocular pain that is markedly reduced following prednisolone plus cyclophosphamide.

Fig. 15. Churg-Strauss syndrome. A 68-year-old man with Churg-Strauss syndrome had periocular pain and restrictions of ocular motility. CT image shows an apparent lateral rectus enlargement.

4.7 Orbital foreign body
Some foreign bodies may cause orbital inflammation. Polymethyl methacrylate (PMMA) is used by cosmetic surgeons in some countries, and can occasionally lead to serious ocular complications including orbital myositis (Figure 16; Kubota & Hirose, 2005; Sato et al., 2007; Silva & Curi, 2004)
Fig. 16. Orbital inflammation associated with a foreign body. Left: A 29-year-old woman underwent cosmetic rhinoplastic surgery by a cosmetic surgeon. She developed ocular pain and a decrease of vision after the PMMA was injected. The right eye was displaced temporally leading to blepharoptosis. Her ocular motility was restricted. Right: T2-weighted MRI shows high signals from the right rectus muscles especially the medial rectus muscle. Reproduced with permission from Kubota T & Hirose H. (2005) Permanent loss of vision following cosmetic rhinoplastic surgery. *Japanese Journal of Ophthalmology*, Vol.49, No.6, pp.535-6, ISSN:0021-5155

### 4.8 Orbital inflammation triggered by drugs
Orbital inflammation including orbital myositis can be caused by bisphosphonates (Fraunfelder & Fraunfelder, 2003; Philips & Newman, 2008; Seth, et al., 2009; Sharma et al., 2008; Subramanian et al., 2003) and also by influenza vaccine (Thurairajan et al., 1997). Physicians should aware of these rare orbital conditions.

### 4.9 Arteriovenous shunting
Carotid cavernous fistulas leads to diffuse symmetric enlargement of most of the extraocular muscles and an enlargement of the superior ophthalmic vein (Figure 17).

Fig. 17. Carotid cavernous fistulas. External photographs of a 65-year-old woman with swelling of the right eyelid and conjunctival injection. CT and MRI images show enlargements of the right superior, inferior, and medial rectus muscles and superior oblique muscles. In addition, an enlargement of the superior ophthalmic vein can also be seen (arrow).

### 4.10 Intramuscular hemangioma of the extraocular muscles.
Intramuscular hemangioma of the extraocular muscles is a rare clinical entity. Patients with intramuscular hemangioma of the extraocular muscles have isolated enlargement of extraocular muscles without pain and ocular motility restrictions (Kiratli et al., 2003). MR images show isointense T1-weight and hyperintense on T2-weight images, compared with extraocular muscles (Kiratli et al., 2003).
5. Treatments

Idiopathic orbital myositis is characterized by a variable natural course of evolution with spontaneous remission (Kubota & Kano, 2007; Slavin et al., 1982) to a corticosteroids-resistant progressive course. The first-line treatment is corticosteroids. It is believed that delayed diagnosis and treatment may lead to permanent dysfunction and both prompt therapy and a slow prolonged steroid taper can prevent ocular motility restrictions and recurrences (Costa et al., 2009; Scott & Siatkowski, 1997). In corticosteroids-resistant cases, the second-line treatment is done in a stepwise manner: first radiation, second immunosuppressive, and third biological agents.

5.1 Steroid treatment

It has been reported that oral NSAIDs are effective for idiopathic orbital myositis in a case series study (Mannor et al., 1997). However, in general the first-line treatment is corticosteroids. In the literature review, different initial doses ranging from 20 to 120 mg a day of oral prednisolone and 1000 mg intravenous methylprednisolone for 3 days have been used, and idiopathic orbital myositis frequently shifts toward recurrent and chronic course up to 75% despite the corticosteroids treatments (Costa et al., 2009).

5.2 Radiation therapy

Radiation therapy ranging from 16 to 30 Gy has been used for orbital myositis in corticosteroids-resistant cases or has been used as a corticosteroid-sparing method. In a review of radiation therapies patients with orbital myositis, approximately one-half of patients had a recurrence (Isobe et al., 2004).

5.3 Immunosuppressive therapy

Several case reports of immunosuppressive therapy for idiopathic orbital myositis have been published. Cyclosporine (Sanchez-Roman, et al. 1993), cyclophosphamide (Gunalp et al., 1996), and methotrexate (Kubota & Kano, 2007) have been use with variable results.

5.4 Biological agents

Therapy with tumor necrosis factor (TNF)-alpha inhibitor, such as infliximab is efficacious for immune-mediated inflammatory conditions, including rheumatoid arthritis and Crohn disease. Several published data showed that biological agents were successful in treating orbital myositis refractory to corticosteroids (Garrity et al., 2004; Miquel et al., 2008; Sahin et al., 2009) including orbital inflammatory disease (Kapadia & Rubin, 2006).

6. Conclusions

The etiology of orbital myositis is unknown. Orbital myositis associated with specific autoimmune disorders especially of giant cell myocarditis and Crohn disease may suggest a clue of pathogenesis of idiopathic orbital myositis, although the incidence of orbital myositis associated with autoimmune diseases is extremely low. Published case series studies have provided the best treatments by different initial doses of corticosteroids. But it appears to be difficult to evaluate the effectiveness for them using a meta-analysis. The acute and chronic/recurrent type of orbital myositis clearly respond differently to corticosteroids, therefore the type should be diagnosed before the treatment. Fat-suppressed T2-weighted

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MR imaging may provide one of the predictive factors. Some orbital myositis refractory to corticosteroids can be effectively treated by immunosuppressive therapy and biologic agents. However, there is no randomized or comparison study and predictive factors for the effectiveness of different treatments.

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The term "myositis" covers a variety of disorders often designated "idiopathic inflammatory myopathies". Although they are rather rare compared to other rheumatic diseases, they often cause severe disability and not infrequently increased mortality. The additional involvement of important internal organs such as the heart and lungs, is not uncommon. Thus, there is a great need for a better understanding of the etiopathogenesis of myositis, which may lead to improved treatment and care for these patients. Major advances regarding research and medical treatment have been made during recent years. Of particular importance is the discovery of the Myositis specific autoantibodies, linking immunological and pathological profiles to distinct clinical disease entities. A wide range of aspects of myopathies is covered in the book presented by highly qualified authors, all internationally known for their expertise on inflammatory muscle diseases. The book covers diagnostic, pathological, immunological and therapeutic aspects of myositis.

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