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Orbital Myositis with Rapid Successful Treatment with Corticosteroids

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

A 41-year-old Japanese female was admitted to our hospital with a history of right abducens nerve palsy, right gaze diplopia, right eye pain and double vision. Thyroid function, thyroid autoantibody levels, and tests for other pathologies were normal. Orbital contrast-enhanced short-TI Inversion Recovery-magnetic resonance imaging before treatment showed contrast-enhanced, severe lateral rectus and rectus superior muscle swelling in the right eye. We therefore diagnosed this patient as orbital myositis. Intravenous glucocorticoid pulse therapy with methylprednisolone (mPSL) was initiated. After prescribing a daily dose of 1,000 mg of mPSL three times a week, all symptoms, including physical abnormalities, disappeared. The patient was discharged on day 14 after hospitalization and was prescribed oral PSL (30 mg/day). This report indicates that early and initial adequate treatment with a high dose of mPSL is very effective for orbital myositis treatment. However, the possibility of recurrences must be always considered at subsequent follow-up.

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Keywords: Orbital myositis; glucocorticoid therapy; orbital contrast-enhanced STIR-magnetic resonance imaging; rapid successful treatment.

1. INTRODUCTION

Orbital myositis is a noninfectious inflammatory condition primarily affecting extraocular muscles. Acute myositis is the second most common reason for nonspecific orbital inflammation [1]. Inflammation of orbital tissue clinically leads to proptosis, unilateral or bilateral eye pain, vision due to restricted eye movement and other symptoms reflecting inflammation such as eyelid swelling, eyelid erythema, conjunctival redness, chemosis and swollen caruncles [2]. Unilateral presentation is typically seen, but bilateral cases are not uncommon. Diagnosis of orbital myositis is often delayed, and up to half of the affected patients are initially treated for assumed orbital infections [3]. The delay of both correct diagnosis and right treatment may be associated with permanent extraocular muscle dysfunction [4]. Therefore, prompt therapy is recommended to provide physical relief, as from pain during the active phase, to preserve the motor function, as well as to prevent both recurrences and disease sequelae [5]. Here, we report an ideal and rare case of orbital myositis, its treatment, and outcome.

2. CASE REPORT

A 41-year-old Japanese female was admitted to our hospital with a 1-month history of right abducens nerve palsy, right gaze diplopia, right eye pain, and double vision. Before she was admitted to our hospital, she was treated by an ophthalmologist at another hospital with oral betamethasone (3 mg/day), as steroid therapy, and vitamin B complex for 7 days; however, her condition did not improve. The patient had a history of iron deficiency anemia. An initial examination on the first visit revealed that the patient was 163.0-cm tall and weighed 48.0 kg, with a body mass index of 18.1. Her blood pressure was 110/65 mmHg, with a regular pulse rate of 82 beats/min and her body temperature was 36.5ºC. Physical examination revealed a horizontal gaze diplopia, bulbar position in abduction, marked limitation of her extraocular movements in the right position of gaze and mild right retro-orbital pain. The patient did not have dizziness, nystagmus, motor dysfunction, or difficulty in swallowing solids or liquids, and her osteotendinous reflexes, bilateral plantar reflexes and speech were normal. Several diagnoses were considered, including thyroid disease, ocular myasthenia gravis, infection, ocular-pharyngeal syndrome and orbital myositis.

The patient's laboratory data are shown in Table 1. Thyroid function and thyroid autoantibody levels were normal. Other biochemical tests revealed the following: acetylcholie receptor antibody (AChRAb) was <0.1 nM/l, HbA1 – 5.4%, and antinuclear antibody, Perinuclear anti-neutorophil cytoplasmic antibody (ANCA), angiotensin converting enzyme (ACE) were all negative, and IgG4 was normal. Examination of cerebrospinal fluid revealed that cell count was 2/mm3 (monocyte), protein concentration was 27 mg/dl, glucose concentration was 57 mg/dl, and Cl concentration – 128 mEq/l, respectively. Orbital contrast-enhanced short-TI Inversion Recovery-magnetic resonance imaging (STIR-MRI) before treatment showed contrast-enhanced, severe lateral rectus and rectus superior muscle swelling in the right eye (Figs. 1A, B). Thus, we diagnosed this patient as orbital myositis.

Intravenous glucocorticoid pulse therapy with methylprednisolone (mPSL) was initiated. A daily dose of 1,000 mg of mPSL was prescribed three times a week, followed by oral PSL (40 mg/day) for 10 days. After the treatment by mPSL (3 days), all symptoms and physical abnormalities had disappeared rapidly and the patient recovered without recurrence. On day 14, orbital contrast-enhanced STIR-MRI showed either disappearance or remarkable decrease in the previously observed abnormalities (Figs. 1C, 1D) and the patient was therefore discharged with a prescription of oral PSL (30 mg/day). After she was discharged from the hospital, oral outpatient care of PSL was then gradually decreased by 10mg/week up to no prescription. Although it have taken more than about one month since discharge, she have not recurred with no medication yet.

3. DISCUSSION

Orbital myositis is described to be extremely sensitive to corticosteroid therapy. For single muscle disease, treatment with nonsteroidal antiinflammatories or mPSL at a dose of 20 mg is usually recommended [6]. However, in initial inadequate treatment regiments, a risk of recurrence is possible [7]. In this case, a low-dose oral steroidal treatment prior to admission

at our hospital proved ineffective. We therefore decided that an initial adequate treatment with mPSL was required. Early aggressive treatment of orbital myositis may reduce the degree of fibrosis and late cicatricle sequelae. The response to treatment is usually fast, as well as the current case, occurring within days of initiation, with initial clinical success in at least 67% of the treated patients [8]. In the presented study, the response to treatment was unusually fast; after three days of treatment with intravenous glucocorticoid pulse therapy with mPSL, all symptoms, including physical abnormalities, disappeared. We speculate that one of the factors contributing to this response was the short time from disease onset to diagnosis and treatment. A good response to corticosteroid therapy, however, is not diagnostic of orbital myositis because many other disorders, such as lymphoid tumors, orbital sarcoidosis, granulomatosis with polyangiitis, Tolosa–Hunt syndrome, IgG4-related disease, and thyroid-related eye disease can show prompt and transient clinical improvement in application of this therapy [9–11]. Therefore, thorough investigation before treatment is important for the diagnosis by clinical and imaging characteristics. The orbital myositis changes could be best seen on contrast-enhanced STIR-MRI fat suppressed post-gadolinium scans. Coronal images are the best for assessing muscle belly size, with axial and sagittal views used to assess tendon involvement. These sequences are edema and the fat suppression processing allows a clearer visualization of the structures within the orbital fat [12]. The contrast-enhanced STIR-MRI is also very useful in the patient diagnosis.

Recurrences can present in any extraocular muscle on either side. In the literature, 52%−55% of patients experience recurrences, which requires corticosteroid therapy and approximately 20% require adjuvant radiotherapy [13,14]. There might be multiple recurrences that may require repeated corticosteroid therapy. Some clinicians consider that a prolonged tapering off of steroids is preferable to reduce the frequency of recurrences, although there have been no randomized prospective trials to define the best practices for the therapy of that disease. In spite of the modality of treatment, some patients experience persistent motor dysfunction in one or more fields of gaze, even after good outcome of the acute inflammatory phase. According some researchers, aggressive and early disease therapy might be able to minimize these types of sequelae [15,16]. We also agree with this. An immune-mediated mechanism is thought to be or form the base for the process of orbital myositis; however, the exact pathogenesis remains unknown. The autoimmune nature of the disease underlies the rationale for using steroids, considering that the beneficial effects of this group of drugs have been observed in other autoimmune diseases. Although the mechanisms of action of steroids in patients with orbital myositis are poorly understood, we consider that the effects on the activation of helper T cells and the proliferation of B cells, activated T cells, and antigen-presenting cells might play a role. We suggest that one of the reasons for the clinical effects of high-dose steroid treatment is the blockage of pathways by which immune activation causes cellular damage and undesirable clinical outcomes at a stretch. Although pathology among patients with recurrent palsy of extraocular muscles is an uncommon, a possible diagnosis of orbital myositis must always be considered.

Table 1. Laboratory data

Blood count
WBC 5380 /μL (Lymphocyte 12.3%, Neutrophil 77.1%), RBC 470 × 10^4/μL, Hb 9.0 g/dL, Hct 30.9 %, MCV 66 fl, MCH 19.1 pg, MCHC 29.1 %, Plt 21.2 × 10^4/μL.
Blood chemistry
TP 7.1 g/dL, Alb 4.6 g/dL, AST 12 IU/L, ALT 8 IU/L, ALP 200 IU/L, CPK 54 IU/L, Cre 0.59 mg/dL, T-Chol 124 mg/dL, glucose 115 mg/dL, Na 140 mEq/L, K 3.7 mEq/L, Cl 107 mEq/L, Ca 8.8 mg/dL, Fe 10μg/dl, UIBC 417μg/d, CRP 0.02 mg/dl, ESR 9 mm/h, HbA1 5.4%.
Other findings
TSH 0.60μIU/mL, FT3 2.37 pg/mL, FT4 1.17 ng/dL, TRAb < 0.1 IU/ml, TgAb 14 IU/mL, TPOAb 6.0 IU/mL, Thyroglobulin (-), Microsome (-), AChRAb < 0.1 nM/l, ANA <40 times, ACE 9.3 U/L, MPO-ANCA < 0.1 U/mL, PR3-ANCA <0.1 U/mL, IgG4 21.3 mg/dl.

ESR; erythrocyte sedimentation rate, CRP; C-reactive protein, TSH; thyroid-stimulating hormone, FT; free triiodothyronine, TRAb; TSH receptor antibody, TPOAb; thyroid peroxidase antibody, AChRAb; acetylcholie receptor antibody, ANA; antinuclear antibody, ACE; angiotensin I converting enzyme, ANCA; anti-neutrophil cytoplasmic antibody

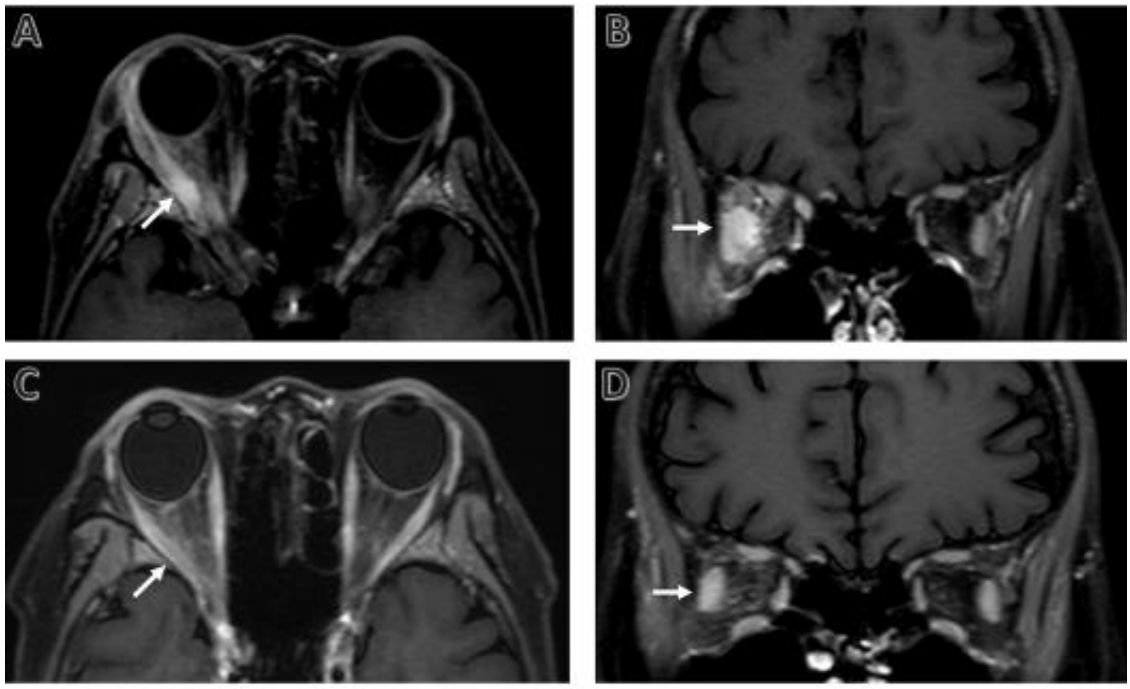

Fig. 1. Orbital contrast-enhanced STIR- MRI. (A) Axial fat-suppressed contrast-enhanced T1 shows severe lateral rectus muscle swelling, contrast enhancement in right eye. (B) Coronal fat-suppressed contrast-enhanced T1 shows severe lateral rectus muscle swelling, contrast enhancement and rectus superior muscle swelling in right eye. (C) and (D): On 14 day; (C) Axial fat-suppressed contrast-enhanced T1 shows the disappearance or remarkable decrease of the abnormalities in right eye. (D) Coronal fat-suppressed contrast-enhanced T1 shows the disappearance or remarkable decrease of the abnormalities in lateral rectus muscle. Arrows indicate lateral rectus muscle in right eye

4. CONCLUSION

The presented report indicates that early and initial adequate treatment with high-dose of mPSL is very effective. In addition, contrast-enhanced STIR-MRI is very useful for diagnosis of orbital myositis. However, the possibility of recurrences must be always considered during subsequent follow-up.

CONSENT

The responsible for the patient gave informed consent for the case report to be published.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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