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A New Onset Recurrent Myositis Idiopathic Orbital Inflammation in Adult Male Patient : A Case Report

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Abstract:Idiopathic orbital inflammation (IOI) is a benign, non-infective clinical syndrome characterized by inflammation of the orbit without identifiable local or systemic disorders. Orbital myositis is one classification of IOI and rare in adult male patients. In this article, we report a 41-years-old male who presented to the eye clinic with swollen, redness, pain, chemosis, and squints on his left eye 2 weeks ago. He had a history of recurrent swollen on his left eyelid and self-treated with methylprednisolone. Visual acuity on right and left eye were 20/25 and 20/30, respectively. The intraocular pressure on the right and left eyes were 18 mmHg and 24 mmHg, respectively. No abnormality in the fundus examination. There was a restriction on the left eye movement to all gaze, and the eye was fixed to the superior temporal direction. Head MRI-scan showed hypertrophy of the lateral and medial rectus muscles on the left eye without any other abnormality in the intracranial cavity. The laboratory examination showed leukocytosis, neutrophilia, lymphopenia, normal thyroid function, low C-Reactive Protein level, and a negative result for the antinuclear antibody test and anti-ds-DNA test. The patient was diagnosed with myositis of idiopathic orbital inflammation and was treated with a high dose of methylprednisolone 1 mg/kg BW/day. Two weeks later, the patient had improvement in his eye movement and the swollen was resolved. The treatment is still ongoing. Finally, we can conclude that comprehensive evaluation and treatment with high-dose corticosteroid results in a better outcome in an adult male patient with myositis IOI.

***Keywords:*Case report, idiopathic, male, myositis, orbital inflammation**

I. INTRODUCTION

Idiopathic orbital inflammation (IOI) is a benign, non-infective clinical syndrome characterized by inflammation of the orbit without identifiable local or systemic disorders. The exact etiology is unknown, but infection and immune-mediated processes have been postulated. IOI accounts for approximately 8%-10% of all orbital mass lesions. It is the third most common orbital disease after thyroid orbitopathy and lymphoproliferative disease. In adults, IOI tends to be unilateral, but children more commonly present with bilateral disease. Clinical presentation usually is an acute onset of pain, redness, chemosis, proptosis, and periorbital edema. Decreased vision and diplopia can also occur based on the orbital structure involved.[1–3]

There is no universally accepted classification for IOI. Depending on the orbital site of involvement, IOI can be divided into anterior, diffuse, apical or posterior, myositis, and dacryoadenitis. IOI is usually seen in the fifth decade and there is no sex predilection. However, orbital myositis less commonly affects adult men with a prevalence rate of 43%.[4] This study aims to characterize the clinical features, radiological evaluation, laboratory findings, and treatment outcomes in the case of myositis IOI in an adult male patient who had a complete resolution with corticosteroid therapy.

II. CASE PRESENTATION

A 41-year-old male presented to the eye clinic with swollen, redness, and pain in his left eye 2 weeks before. He complained of squint on his left eye without any proptosis, visual disturbance, headache, or fever. He had a history of squint, recurrent swollen, and mild ptosis on his left eyelid. Then he had been diagnosed with IOI and treated with methylprednisolone orally. But the patient self-treated with methylprednisolone 8 mg bid orally for more than 5 years in case the symptoms appear. No history of diabetes mellitus, hypertension, trauma, fever, or autoimmune disease before.

The patient was conscious with blood pressure 126/97 mmHg. Other vital signs were normal. The eyelid of the left eye was edema and tender with chemosis in the conjunctiva. Visual acuity on right and left eye were 20/25 and 20/30, respectively. The intraocular pressure on the right and left eyes were 18 mmHg and 24 mmHg, respectively. There was no abnormality in the fundus examination. There was a restriction on the eye movement of the left eye to all gaze (-3 in the lower medial quadrant, -2 in the upper medial quadrant, and -1 in the lateral quadrant) and the eye was fixed on the superior temporal direction (Figure 1). No other neurological deficit was found.

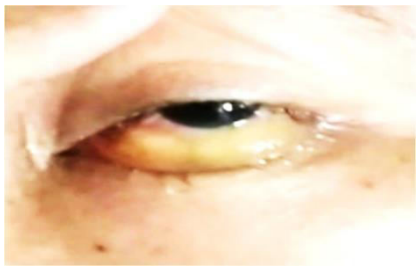

Figure 1. Anterior segment of the left eye showing edema and tenderness of the left eye.

Initially, we diagnosed the patient with orbital cellulitis, thyroid eye disease, or metastatic carcinoma. The patient was referred for Head MRI-scan without contrast, complete blood count, and thyroid function test to established the diagnosis. Head MRI-scan showed a homogenous enhancement lesion on medial and lateral rectus muscles of the left eye that indicated an inflammatory lesion on extraocular muscles. No other abnormality was found in the intracranial cavity (Figure 2). The laboratory examination showed marked leukocytosis (13.600/ µL), neutrophilia (85.7%), lymphopenia (9.6%), and normal thyroid function (TSH 0.848 uIU/ mL and total T4 level (5.36 ug/dL). Based on these results, we diagnosed the patient with orbital myositis.

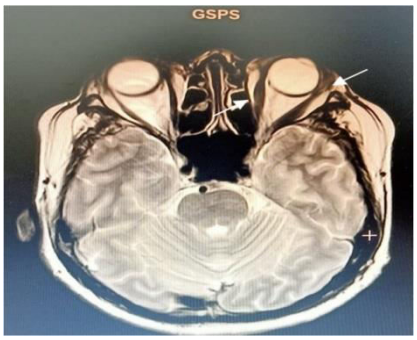

Figure 2. Head MRI-Scan without contrast of the patient in axial view showed a homogenous enhancement lesion on medial and lateral rectus muscles of the left eye that indicated an inflammatory lesion on extraocular muscles (white arrow).

He was then suggested to have further laboratory examination to ruled out the etiology, such as the systemic inflammatory state. The results revealed a normal inflammation marker (hs CRP 1.50 ng/L) and a

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negative result for systemic lupus erythematosus markers (ANA Test 0.50 AU/mL CLIA and Anti Ds DNA 0.50 IU/mL).

The patient has diagnosed with myositis idiopathic orbital inflammation and was started with oral methylprednisolone 1 mg/kg BW / day. Two weeks later, the ocular movement and swelling were partially resolved (Figure 3). The patient also was referred to the internal medicine department then treated with levofloxacin 500 mg once daily and lansoprazole 30 mg twice daily. The patient was on a maintenance dose of steroids and was kept under observation with monthly follow-up to prevent local recurrence or disease progression.

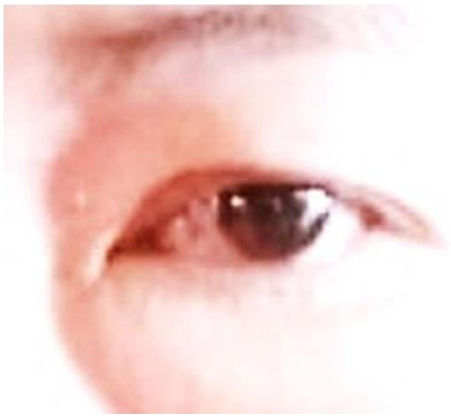

Figure 3. Anterior segment of the left eye showed improvement of the symptom.

III. DISCUSSION

IOI may present in an acute, subacute, or chronic fashion. It is usually unilateral, but can also bilateral disease. IOI is usually seen in the fifth decade and there is no sex predilection. However, orbital myositis most commonly affects young adults in the third to the fourth decade of life and shows a female predilection.[3,4] In this case, the patient was a male with unilateral involvement and showed a less common cause. There were no clinically difference between male and female patients, despite less preceded with history of upper respiratory tract infections and more subacutely in male patients.[4]

IOI may present with a wide range of clinical manifestations. There is no universally accepted classification for IOI and can be conveniently divided into categories including anterior, diffuse, apical or posterior, myositis, and dacryoadenitis. Other rare IOI types include periscleritis, perineuritis, and focal mass. Different clinical manifestations and radiological examinations can help to distinguish among them.[3,5]

This patient presented with swollen, redness, pain, chemosis, and squint on his left eye. There was a restriction on the left eye movement to all gaze, and the eye was fixed to the superior temporal direction. Based on the literature, the anterior IOI is frequently characterized by pain, periorbital swelling, conjunctival chemosis, and limited ocular motility. The diffuse IOI is frequently presented with proptosis and similar clinical features but more severe than anterior IOI. Apical or posterior IOI is associated with poorer visual outcomes and can also present with orbital pain, restricted eye movement, minimal proptosis, and intracranial extension indicated with a unilateral headache. Myositis IOI involves single or multiple extraocular muscles and is frequently presented with unilateral orbital or periorbital pain, diplopia, ocular motility restriction, proptosis, eye swelling, and conjunctival injection. Myositis IOI most frequently involved muscle in the medial rectus followed by the superior, lateral, and inferior rectus muscles. Dacryoadenitis IOI is characterized by acute painful, firm, erythematous mass with edema in the lateral upper eyelid, and S-shaped ptosis sometimes associated with dry eye.[3,6] Based on clinical presentation, we suspected the patient with anterior IOI or myositis IOI. To distinguish between them, we performed a radiological examination.

Orbital and systemic diseases that may present with similar signs and symptoms include orbital cellulitis, auto-immune disorders (ie. systemic lupus erythematosus, rheumatoid arthritis), thyroid eye disease, malignancy (ie. Lymphoma, metastatic carcinoma, leukemia), IgG4-related orbital disease, or Sarcoidosis.[7] In this patient, laboratory examination and MRI was done to exclude other mimic diseases. MRI is the modality of choice for the evaluation of orbital inflammation because of its superior soft-tissue contrast and spatial resolution, as well as it's possible to generate functional images such as diffusion-weighted imaging (DWI) and perfusion-weighted imaging (PWI). In this patient, the head MRI scan was done to eliminate the possibility of thyroid eye disease, sinusitis (one of the causes of orbital cellulitis), and metastatic carcinoma. The result showed a homogenous enhancement lesion on the medial and lateral rectus muscles of the left eye. MRI scan can distinguish between the inflammatory lesion and tumor lesion. Inflammatory lesions tend to show a more homogenous enhancement pattern, while tumors and infections will be more heterogeneous due to the presence of non-enhancing components such as necrosis and pus.[5]

Myositis is characterized by a unilateral thickening of one or more extraocular muscles often involving the myotendinous junction with a fusiform configuration on MRI. There may be ill-defined infiltrates throughout the surrounding orbital fat.[8] These are important features in distinguishing myositis from thyroid orbitopathy, which affects extraocular muscles bilaterally and spares the myotendinous junction with an increase in orbital fat volume.[3] Involvement of the perimuscular tendon in recent reports suggests a myositis rather than thyroid orbitopathy which is to enlarge the muscle belly alone.[8] This result confirmed the diagnosis of the patient with myositis IOI. Further thyroid eye disease examinations were not performed because the thyroid eye disease was excluded.

Thyroid function test is the initial laboratory testing for myositis IOI because it is necessary to exclude thyroid orbitopathy, the main differential diagnoses for myositis IOI.[3,6] In this patient, the thyroid function test within normal limits, and according to the result of the head MRI scan, we can exclude the diagnosis of thyroid orbitopathy. So, further thyroid orbitopathy examinations were not performed in this patient.

Another laboratory testing was performed, including complete blood count, erythrocyte sedimentation rate, C-reactive protein, antinuclear antibodies, and anti-Ds-DNA. The result was within normal limits except complete blood count, so we can exclude the diagnosis of systemic lupus erythematosus and orbital cellulitis. Laboratory examination revealed leucocytosis with neutrophilia and lymphopenia that may indicate infections, stressful conditions, chronic inflammation, medication use, bone marrow stimulation, splenectomy, or congenital abnormality.[9] Infection must be considered in this case because there is leucocytosis. The orbit may be the site of specific infections, most commonly by paranasal sinusitis or contiguous spread from infections of the face, teeth, a penetrating foreign body in cases of trauma, and septicemia. Infection most commonly begins in the ethmoid sinus, with the extension of the inflammatory infiltrate into the orbital space. In mild cases, the process may be limited to the preseptal space.[10] In this patient on MRI the paranasal sinus, intraconal and extraconal space was within normal limit; and there was no history of trauma.

Antineutrophil cytoplasmicantibodies, angiotensin-converting enzyme level, rapid plasma regain test, rheumatoid factor, serum IgG4 level, etc are the other valuable laboratory testing.[6,11] It is important to be mindful of atypical micro-organisms, including fungal infections such as mucormycosis and aspergillosis, in patients with diabetes and immunocompromised patients.[10] A histological examination from orbital muscle must be ruled out to exclude infection as the etiology.[12] Also, systemic examinations like urinalysis or another panel should be performed to identify another source of infection. These are our limitations in this case.

Another measurement was normal that suggested the ocular inflammation of this patient likely due to an idiopathic condition. Finally, based on clinical, radiological, and laboratory findings, we diagnosed this patient with myositis idiopathic ocular inflammation.

Generally, the mainstay treatment for IOI is corticosteroid.[3,4] For myositis IOI, corticosteroid dosing usually consists of oral prednisone 1 mg/kg/day for 1-2 weeks followed by a 6-12 week tapered-off. Induction therapy with intravenous methylprednisolone 1000 mg/day has also been utilized to good effect.

Some advocate for lower doses (20 mg oral prednisone per day) in cases with single muscle involvement. Up to 80% of patients treated with steroids alone eventually experience a relapse of myositis during steroid taper and thus steroid-sparing medications are often necessary, such as immunosuppressive agents (ie. methotrexate, cyclosporine,cyclophosphamide, etc.), biologic agents (ie. infliximab, daclizumab, rituximab, and adalimumab), radiation therapy.[13]In this patient, the main treatment with methylprednisolone orally 1 mg/kg/day for 2 weeks and tapered off in the next 4-6 weeks gave an improvement of the symptom. In line with the study by Ota et al (2015), giving methylprednisolone as early as possible at a dose of 60 mg/day followed by tapering-off 10 mg/week can provide complete resolution. Improvement by methylprednisolone can be achieved even in the form of single therapy, compared to prednisone which requires combination therapy with cyclosporine to achieve significant improvement.[14]

In myositis IOI patients, complete symptom relief show in 63% of patients. However, 35% of patients experience only partial relief and are left with persistent motility dysfunction, pain, or visual loss. In 2% of patients, treatment is ineffective. Of the patients who experience relief, one-third have a recurrence of disease at the same or a different location.[6]The frequency of recurrence about six times and can occurred in 81,8% of the patients. Improper dosage or duration of corticosteroid treatment may underlie the recurrent episodes.[4]In this patient, the recurrences may be due to inadequate dosage of self-treatment with methylprednisolone before.

The patient felt satisfied with the treatment given. The patient felt that there are many improvements in his eyes compared to when he first came. Until the last control, the patient did not complain of swelling in his eyes anymore.

Limited of this case is we did not perform laboratory testing for angiotensin-converting enzyme level, rapid plasma regains test, rheumatoid factor, and another test (ie. urinalysis, etc.) to deeply investigate the cause of the disease,[6,7,12]and rule out the infection process according to laboratory results in this patient. Comprehensive evaluation and initial treatment with high-dose corticosteroid results in a better outcome in myositis IOI patients.

IV. CONCLUSION

Myositis idiopathic orbital inflammation is a rare case in adult male patients. A comprehensive evaluation includes careful history taking, ophthalmological examination, radiologic examination, and a large laboratory test are needed in diagnosed the disease. The main treatment with high-dose corticosteroid and tape-off can result in complete resolution of the symptom.

V. ACKNOWLEDGMENT

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