A patient with diplopia and upper eyelid edema: a case report

Igor Šivec Trampuž, Dragica Kosec

Department of Ophthalmology, University Medical Centre Ljubljana, Ljubljana

Correspondence:
Igor Šivec Trampuž,
e: sivec.igor@gmail.com

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Abstract

Idiopathic orbital inflammatory disease, previously referred to as orbital pseudotumor, is a non-neoplastic, non-infective disease. The inflammatory process can involve one or more orbital soft tissues; such as extraocular muscles, tear gland, scleral or episcleral tissue or orbital fat. Orbital myositis typically presents with a sudden onset of restricted ocular motility, diplopia, congestive proptosis, eyelid ptosis, periocular swelling and conjunctival hyperemia. A thorough workup is essential for ruling out other entities.

This case report presents a 47-year-old man with a sudden onset of right upper eyelid swelling and diplopia. He had no previous health problems. Initial ophthalmologic workup and ultrasound findings suggested an orbital mass. A MRI and biopsy of the mass confirmed the diagnosis of isolated superior oblique muscle myositis. The patients was treated with systemic steroids. He showed a good response and fast regression. During two years' follow-up no recurrence was noted.

Isolated superior oblique muscle myositis is extremely rare. We present one of the eleven cases documented between 1988 and 2015.

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1. A case report

A 41-year-old patient was admitted to the University Eye Hospital due to a sudden onset of double vision and right upper eyelid swelling. A week before the patient noticed swelling of the upper eyelid of his right eye and double vision that worsened when looking to the left and upwards. Since then he has been in good health and had no eye problems. Ten years previously he had had probing and irrigation of the lacrymation system due to tearing. There was no relevant information in his past medical, social and family history, or traveling abroad; he denied previous injury or surgery. He had an allergy to peniciline.

At initial presentation, his corrected visual acuity was 1.0, intraocular pressure was 13 mmHg and 15 mmHg for the right and left eye respectively. Color perception testing (Ishihara) was normal. Pupilary responses were normal. Mildly painful and restricted elevation in adduction and upper eyelid edema in his right eye were observed. There was no »clicking«, crepitation or mass felt on palpation. Biomicroscopy and fundus examination were unremarkable. The Goldmann visual

field test was normal. Brief general physical and neurological examination were unremarkable.

Orbit ultrasound showed a low-reflective area in the anterior upper nasal segment of his right orbit (6 × 8 × 12 mm). There were no pathological changes in his left orbit.

A MRI showed a well demarcated pathologic formation along the superior oblique muscle that was pushing away the superior rectus muscle. The formation was isointense to muscle on T1 sequence and hyperintense on T2 sequence. A contrast-enhanced MRI showed homogeneous hyperintensity and enhancement of the superior oblique muscle (Figure 1).

The differential diagnosis included lymphoproliferative, metastatic and other neoplastic masses, myositis, thyroid orbitopathy and less likely sarcoid, autoimmune connective tissue diseases and parasitic infections.

Laboratory tests, including C-reactive protein (CRP), sedimentation rate (SR), thyroid stimulating hormone (TSH), other thyroid hormones (T3, T4), complete and differential blood counts were negative.

Biopsy of the suspected mass was performed. Histopathology revealed mild inflammatory changes that could have been the consequence of orbital myositis. There were no cancerous or granulomatous cells (Figure 2).

Based on the results the patient was started on oral corticosteroid therapy with an algorythm of methylprednisolone 1 mg/kg of body-weight per day for three consecutive days, and a slow taper of 4 mg every three days. In addition, the patient received omeprazole for gastric protection. One day after initiating the therapy a dramatic improvement occurred.

On follow-up exams the patient showed no recurrence of symptoms. A follow up MRI (Figure 3) showed slight enlargement of the superior obliqe muscle. There were no signs of recurrence in the next two years' follow-up.

2. Discussion

Idiopathic orbital myositis is a subgroup of idiopathic orbital inflammatory syndrome (IOIS), formerly termed pseudotumor. Orbital myositis is a rare inflammatory disease that affects single or multiple extraocular muscles. In a series of 75 patients studied by Siatkowski and collegues, 68 % had single muscle involvement, in 22 % two muscles were involved and in 10 % three or more muscles were affected. Lateral and medial recti were most frequently affected (33 % and 29 %, respectively), followed by the superior rectus (23 %), the inferior rectus

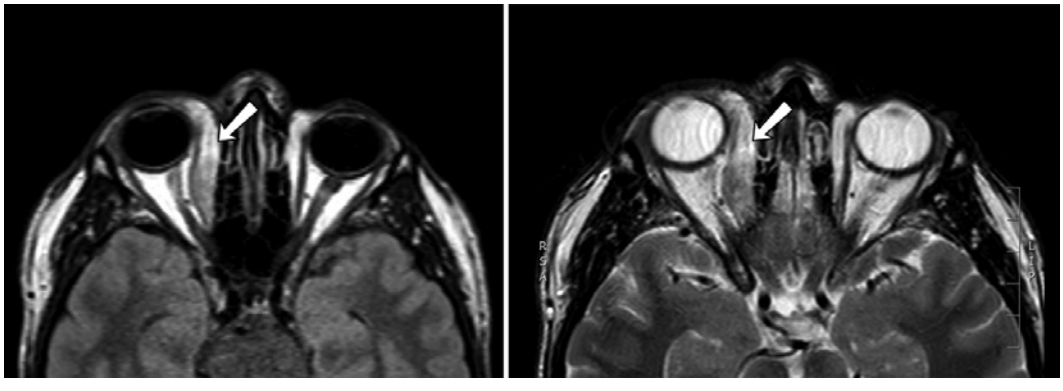


Figure 1: A MRI showing enlargement of the superior oblique muscle.

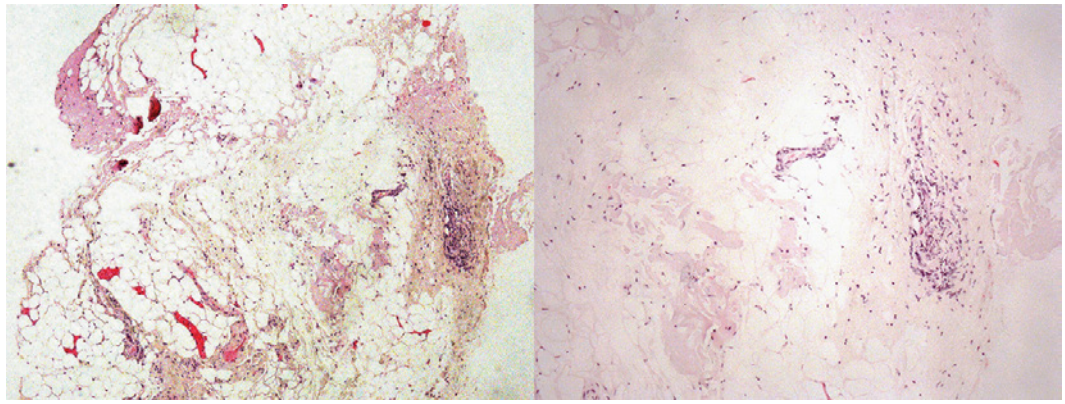


Figure 2: Histopathologic specimen with mild inflammatory changes.

(10 %), the inferior obliqe (3 %) and the superior oblique muscle (2 %) (1-3,14).

To our knowledge, only 11 cases of isolated superior oblique muscle involvement, including this case (Figure 4), were documented between 1988 and 2015. One of the patients has been confirmed to have Wegener's granulomatosis (Salam, 2008) and in two cases relevant information was lacking (Sekhar 1993 and Muralidhar 2015) (1,2,4,11).

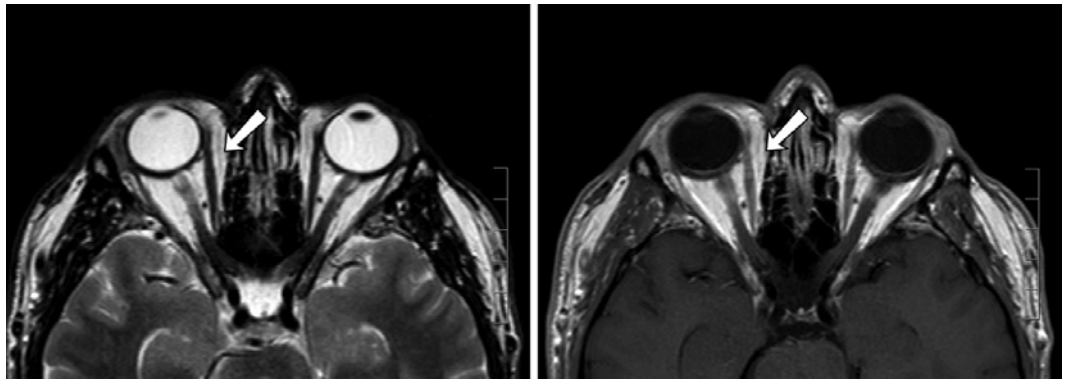
Idiopathic orbital myositis typically presents with a sudden onset of eye movement-evoked pain (95 %), double vision (85 %), conjunctival hyperemia (70 %), proptosis (60 %), eyelid swelling and periorbital edema. Visual acuity is generally unaffected (1-3,6,12-14).

Orbital myositis must be differentiated from several diseases; such as thyroid-associated orbitopathy, neoplasms, cellulitis, sarcoidosis, foreign body granuloma.. Imaging, laboratory tests, and sometimes biopsy, are necessary (6,7,9,12).

Thyroid-associated orbitopathy is an autoimmune disease associated with thyroid disease, mediated by circulating stimulating autoantibodies. It is the most common cause of orbital disease. The onset is characteristically subacute or chronic rather than acute. Bilateral involvement is very common. The most commonly affected muscles are the inferior and lateral recti. Smoking is closely related to the severity of symptoms (5,6,8,12).

Orbital cellulitis is a bacterial infection. It has an acute onset and a progressive development of signs and symptoms. Even though its signs and symptoms are sometimes similar to those of orbital myositis, imaging studies may help to differentiate between the two entities (7,9,12).

Primary and metastatic tumors may mimic orbital myositis (atypical myositis). Imaging studies or biopsy can be used to differentiate between them (7,9,12).



Slika 3: A MRI showing a small residual enlargement of the superior oblique muscle.

Table 1: Overview of the documented cases of isolated superior oblique muscle involvement.

Study	Age	Sex	Involved muscle	Presentation	Duration of symptoms	Orbital imaging	Systemic findings	Treatment	Follow up
Lee Wan (1988)	19	M	Left SO	Pain, periorbital oedema, ophthalmoplegia, conjunctival injection, proptosis	7 days	CT: a irregular mass along the Left superonasal orbit; Echography: low-reflective enlargement of SO muscle and tendon	afebrile leucocytosis	Initially antibiotics, oral steroids	After 3 weeks, persistent Brown's syndrome
Sekhar (1993)	NP	NP	Right SO	A mass in the upper medial orbit	NS	CT: enlarged SO muscle	NS	NS	Loss to follow-up
Moorman (1995)	31	Ž	Left SO	Pain, diplopia, headache	35 days	CT: swelling of the insertion and belly of SO muscle Echography: reduced reflectivity of the belly and swelling of the insertion of SO	NP	Oral steroids	After 6 months, no recurrence
Stidham (1998)	57	M	Right SO	Pain, swelling, diplopia, proptosis, mild ptosis	5 days	CT: enlarged SO muscle without tendon involvement	NP	Oral steroids	After 5 months, no recurrence, residual restriction of elevation
Tsai (2006)	33	F	Right SO	Pain, swelling, diplopia, conjunctival injection	28 days	CT: enlarged SO muscle without tendon involvement MRI: a fusiform swelling of SO muscle, and enhanced heterogeneously with gadolinium	NP	Pulse steroids	After 12 months, no recurrence
Levine (2007)	60	Ž	Right SO	Pain, diplopia, proptosis	NS	CT: enlarged SO muscle	NP	Oral steroids	NS
Salam (2008)	66	Ž	Right SO	Pain, diplopia, ophthalmoplegia	6 months	MRI: enlarged SO muscle	elevated cANCA	Oral steroids and immuno-suppresants	WG
Fleischmann (2012)	34	Ž	Right SO	Pain, diplopia, headache	10 days	MRI: enlarged SO muscle	NP	oral steroids	After 2 weeks full recovery
Han (2014)	57	M	Left SO	Pain, diplopia, headache	3 months	CT: hypertrophied SO muscle	thyroid nodule	oral steroids	After 4 months, no recurrence
Muralidhar (2015)	9	Ž	Right SO	Pain, diplopia	2 days	CT: enlarged SO muscle	NP	Oral steroids	Loss to follow up
Kosec	41	M	Right SO	Pain, diplopia, upper eyelid edema	7 days	MRI: enlarged SO muscle	NP	Oral steroids	After 12 months, no recurrence

NS:not stated, CT:computed tomography, MRI:magnetic resonance imaging, SO: m.obliqus superior, WG: Wegeners granulomatosis, NP: nothing particular

Vasculitis associated with anti-neutrophil cytoplasmic antibodies causes similar orbital changes as myositis(e.g. Churg-Strauss syndrome and Wegener's granulomatosis) (4,7,12).

Some foreign bodies may cause late-onset orbital inflammation (7,9,12-14).

In the majority of cases the cause of orbital myositis is unknown (idiopathic orbital myositis), however some cases with known etiology have been reported (4,6,7,9,12).

Spirochetes (Borrelia burgdorferi), viruses (Herpes zoster virus), and bacteria (Group A streptococci) are microbes that can cause infectious orbital myositis. Parasitic infections are more common in developing countries, e.g. cysticercosis in India (6,7,9,12).

Orbital myositis may be associated with relatively specific autoimmune diseases; including systemic vasculitis, Crohn's disease, systemic lupus erythematosus, reumathoid arthrytis, and scleroderma (7,9,12-14).

Our patient had an isolated form of superior oblique muscle myositis presenting with upper eyelid edema, diplopia and mildly painful and restricted elevation in adduction. There was no conjunctival injection or chemosis. The patient had no history of trauma. The results of laboratory tests were negative and ruled out thyroid dysfunction, sarcoidosis, and acute or chronic systemic inflammation. Orbit ultrasound and MRI studies revealed a mass which was not typical of a neoplastic growth, endocrine orbitopathy or cellulitis. A biopsy of the suspicious mass was performed. Histopathology revealed only mild inflammatory changes with no signs of autoimmunity or inflammatory vascular disease.

The goal of treatment is to intercept the systemic inflammatory cascade in order to reduce the patient's discomfort and prevent permanent damage to the orbital contents. The first-line treatment option are oral corticosteroids with a slow taper. In corticosteroid-resistant cases one should consider radiation or immunosuppressive therapy. All patients with documented isolated superior oblique myositis, including our case, responded favourably to systemic steroid treatment (1,2,6,7,10,13).

3. Conclusion

Idiopathic orbital inflammatory disease covers a spectrum of non-neoplastic and non-infective orbital lesions. The inflammatory process may involve any or all of the orbital tissues, e.g. extraocular muscles, lacrimal gland, scleral and episcleral tissue and orbital fat. The most rare muscle to be affected, especially in isolated form, is the superior oblique muscle. The most sensitive diagnostic tool is orbital MRI, however other diagnostic modalities should also be considered.

In our patient, the inflammation subsided and symptoms dramatically improved one day after initiating systemic corticosteroids. There were no signs of recurrence on follow-up examinations in the next two years.

References

1. Kau HC, Kao SC, Peng CH, Hsu WM, Tsai CC. Methylprednisolone pulse therapy in patient with isolated superior oblique myositis. Eye. 2006;20(9):1106–1109.
2. Zielinski K, Levine M. Case study: Superior oblique muscle enlargement presents as orbital myositis. Ocular Surgery News. 2007[cited 2015.12.01]. Available from: https://www.healio.com/ophthalmology/oculoplastics/news/print/ocular-surgery-news/%7B0fe18712-49b1-4a27-9d22-e1a48d4f2837%7D/case-study-superior-oblique-muscle-enlargement-presents-as-orbital-myositis.

3. Siatkowski RM, Capó H, Byrne SF, Gendron EK, Flynn JT, Muñoz M, et al. Clinical and echographic findings in idiopathic orbital myositis. Am J Ophthalmol. 1994;118(3):343-50.
4. Salam A, Meligonis G, Malhotra R. Superior oblique myositis as an early feature of orbital Wegener's granulomatosis. Orbit. 2008;27(3):203-6.
5. Jaki-Mekjavić P, Beltram. Obravnava bolnikov s ščitnično orbitopatijo, upoštevaje priporočila EUGOGO (european group on graves' orbitopathy). Zdrav Vestn Supl. 2010;79:I34–38.
6. Schoser BG. Ocular myositis: diagnostic assessment, differential diagnoses,and therapy of a rare muscle disease - five new cases and review. Clin Ophthalmol. 2007;1(1):37-42.
7. Idiopathic Inflammatory Myopathies–Recent Developments. InTech: 2011 [cited 2015.12.15]. Available from: http://www.intechopen.com/books/idiopathic-inflammatory-myopathies-recent-developments/orbital-myositis.
8. Soeters MR, van Zeijl CJ, Boelen A, Kloos R, Saeed P, Vriesendorp TM, et al. Optimal management of Graves orbitopathy: a multidisciplinary approach. Neth J Med. 2011;69(7):302-8.
9. Costa RM, Dumitrascu OM, Gordon LK. Orbital myositis: diagnosis and management. Curr Allergy Asthma Rep. 2009;9(4):316–23.
10. Dubey A, Eidsness E, Koul R. Idiopathic orbital myositis and review of literature. The Internet Journal of Ophthalmology and Visual Science 2009;8(2):1–5.
11. Muralidhar R, Gautam K, Christopher D, Vidhya N, Ramamurthy D. Isolated superior oblique myositis causing acquired Brown's syndrome. Indian J Ophthalmol. 2015;63(4):340-1.
12. Kanski JJ, Bowling B, Nischal KK, Pearson A. Clinical ophthalmology: a systematic approach. 7th ed. Edinburgh; New York: Elsevier/Saunders; 2011.
13. Gordon LK. Orbital inflammatory disease: a diagnostic and therapeutic challenge. Eye (Lond). 2006 ;20(10):1196-206.
14. Šivec I, Kosec D. Diplopija: praktični pristop k diagnostiki. Zdrav Vestn 2015;84:392–402.