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CASE REPORT

YOUNG WOMAN WITH HEADACHE AND DOUBLE VISION: A RARE CASE REPORT ON ORBITAL PSEUDOTUMOR

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Abstract:

An uncommon inflammatory disorder that affects the orbit of the eye is called orbital pseudotumor. It can present in a variety of clinical ways. Its precise etiology is yet unclear, although an immune-mediated response is thought to be involved. A 35-year-old woman arrived to the neurology clinic with symptoms of double vision and persistent headache. There was a mild protrusion of the left eye from the orbital cavity, the eyeball medially rotated in the primary position, and restriction of left-sided eyeball movement towards the left side with complaints of increasing double vision during the attempt. She had no recent medical history or traumatic experiences. Over the course of three weeks, the initial discomfort had increased. There were no known allergies, however the patient was normotensive. Upon examination, the left eye had flare, slight anterior chamber cell, erythema, and eyelid edema. Magnetic resonance imaging showed involvement of the optic nerve, thickening of the extraocular muscles, and augmentation of orbital soft tissues. Inflammatory markers were high in the laboratory results. An orbital pseudotumor was diagnosed. The patient was treated with Inj Methylprednisolone followed by corticosteroids, her symptoms improved and the inflammatory condition subsided. A thorough approach including imaging scans, laboratory testing, and clinical evaluation is necessary for its diagnosis. The importance of recognizing an uncommon presentation and management of orbital pseudotumor with was highlighted in this case report.

Keywords: *Double Vision, Headache, Orbital Pseudotumor, Idiopathic Orbital Inflammatory Syndrome*

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Introduction:

An uncommon inflammatory disorder that affects the orbit of the eye is called orbital pseudotumor, or idiopathic orbital inflammatory syndrome. Several anatomical sites and components, including as the extraocular muscles, glands, and connective tissues, may be affected by the inflammation linked to orbital pseudotumor. The clinical signs of orbital pseudotumor are varied and include proptosis, ocular discomfort, edema, and visual abnormalities.[1] It is thought to be

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caused by an immune-mediated reaction, while the precise cause is yet unclear. Orbital pseudotumor is difficult to diagnose since it necessitates ruling out other orbital disorders by combinations of clinical examination, laboratory tests, and neuroimaging.[1] The usual course of treatment is systemic corticosteroids, but biological treatments and other immunosuppressive medications have also showed promise[2]. The aim of this case report is to provide a comprehensive clinical case of ocular pseudotumor, emphasizing the difficulties in diagnosis and treatment while discussing thorough the strategy for management.

Case report:

A 35-year-old woman first consulted a primary care physician for double vision and persistent headaches for 3 days. As routine analgesics were not beneficial, she was referred to a neurology clinic and after assessment instructed for admission. Her double vision was more during her left lateral gaze. She described pain in the left side of the head including the left eye and cheek. Her pain was dull aching, moderately severe, and persisting in nature. Conventional analgesicswere partially beneficial initially. There was no trauma to her eye. No redness or watering from the eyes. On examination, her heart rate was 88 bpm, normotensive BP 120/75 mmHg and she was afebrile. There was a mild protrusion of the left eye from the orbital cavity, the eyeball medially rotated in the primary position, and restriction of left-sided eyeball movement towards the left side with complaints of increasing double vision during the attempt. Her past medical history was not significant.

We have organized several routine blood tests: Hb 10.8 gm/dl, ESR 60 mm in 1st hour, WBC count 6500/cumm and normal lymphocyte count, blood glucose, and serum creatinine. Serum TSH 2.1 mIU/L (normal value 0.5 to 5.0 mIU/L) MRI of the brain was unremarkable, however, MRI of orbits revealed swelling and contrast enhancement of the extraocular muscles and lacrimal gland of the left side (Fig.-1). Mild thickening and contrast enhancement noted in the left optic nerve sheath (Fig.-2). We have excluded Graves's ophthalmopathy, cerebral venous sinus thrombosis, Tolosa Hunt syndrome, and retro-orbital space-occupying lesion in this case. The patient was diagnosed with orbital pseudotumor based on the imaging data, laboratory results, and clinical presentation. The presence of orbital inflammation, normal intraocular pressure, swelling and contrast enhancement of the extraocular muscles and lacrimal gland, involvement of the optic nerve sheath, and increased inflammatory markers all supported the diagnosis.

After a consultant review, she was treated with intravenous methylprednisolone for five consecutive days which significantly improved her headache as well as double vision. Subsequently, oral prednisolone was instituted daily as a morning dose of 1 mg/kg body weight for one month.

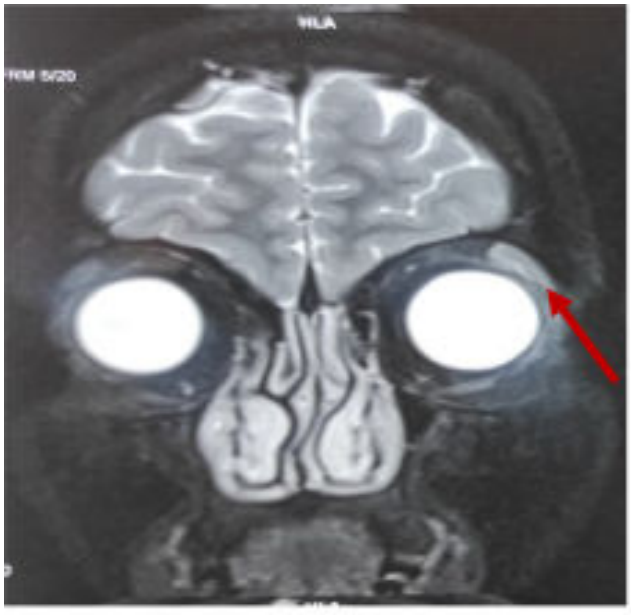

Fig.-1: *MRI orbit (coronal view) showing enlargement and enhancement of left lacrimal gland*

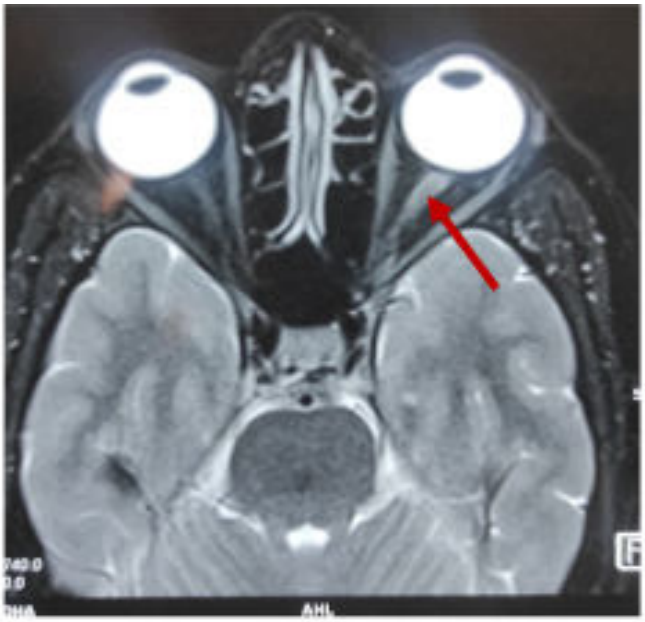

Fig.-2: *MRI orbit (axial view) showing left optic nerve thickening and enhancement*

She was discharged from the hospital after a week, with only oral medication and there was no threat to her vision and safety. There were no symptoms on her follow-up visit after two weeks in the outpatient department. No physical sign was noted. Considering the high recurrence a slow tapering of steroid was adopted. She was instructed regarding the possible side effects of steroids and how to avoid them. A subsequent visit to the outpatient department was scheduled after one month.

Discussion:

Because of its varied clinical appearance and unclear origin, orbital pseudotumor, sometimes referred to as an idiopathic orbital inflammatory syndrome, is an uncommon inflammatory disease that presents diagnostic difficulties.[3] Differentiating orbital pseudotumor from other ocular diseases can be challenging due to the vast range of clinical presentations.[4] Patients most frequently complain eye discomfort, eyelid swelling, diplopia, proptosis, visual abnormalities, and decreased visual acuity. A mix of clinical findings, imaging tests, and laboratory analyses are used to diagnose orbital pseudotumor. Assessing the degree and location of ocular inflammation requires the use of imaging methods such MRI with contrast.[3]

In patients with orbital pseudotumor, it is also quite sensitive in identifying orbital inflammation, which helps distinguish it from other orbital diseases. Laboratory tests that show the existence of systemic inflammation, such as those measuring C-reactive protein and erythrocyte sedimentation rate, operate as supporting measures. However, because they are not specific for orbital pseudotumor, their diagnostic use is restricted.[4]

The main goals of managing orbital pseudotumor are to maintain vision and regulate the inflammatory response.[1,5] Systemic corticosteroids, including prednisone, are thought to be the cornerstone of therapy. Oral corticosteroids at high doses have been shown to quickly reduce inflammation and symptoms in orbital pseudotumor patients.[1]

While it can strike at any age, adults are the ones most affected, with a peak occurrence in the fourth to sixth decades of life.[3] No gender preference is known to exist.[3] It is thought that an immune-mediated response has a role in orbital pseudotumor, albeit the precise cause is yet understood. No definite risk factors or predisposing variables have been identified for orbital pseudotumor.[3] Furthermore, more thorough research is needed to evaluate the long-term effects of various treatment approaches and pinpoint factors that predict treatment response and recurrence.[4]

Nevertheless, prolonged use of corticosteroids is linked to serious adverse effects, which makes the investigation of other therapeutic approaches necessary.[1,2] With encouraging outcomes, immunosuppressive medications including methotrexate and azathioprine have been used as steroid-sparing medicines in recent years.[2,5] Refractory patients have also demonstrated the effectiveness of biologic medicines that target certain inflammatory pathways, such as tumor necrosis factor-alpha inhibitors.[2]

Debate surrounds the function of a biopsy in ocular pseudotumor.[6] When other diagnostic techniques are unable to produce a conclusive diagnosis, or when there is a suspicion of malignancy or unusual presentations, the biopsy may be taken into consideration. However, each person must consider the advantages and disadvantages of performing a biopsy before making the decision.

Conclusion:

Finally, this case report of orbital pseudotumor provides a thorough assessment of a challenging clinical case, emphasizing the diagnostic challenges and therapeutic techniques associated with these uncommon inflammatory diseases. Numerous treatment plans are available due to the disease's varied nature and the development of immuno-suppressive medications. According to retrospective research, individuals often see a reduction in their symptoms. Steroids are often regarded as the first line of therapy for this disease.

Conflict of Interest:

The authors stated that there is no conflict of interest in this study

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This research received no external funding.

Consent:

For the purpose of publishing this case report and any related photos, the parents are written informed consent was acquired.

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