

Orbital pseudotumour presenting as orbital cellulitis

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ABSTRACT

A 33-year-old woman presented to a community emergency department with a 4-day history of monocular orbital pain, photophobia and pain on extraocular movement. Findings included chemosis, conjunctival injection and restricted extraocular movements causing strabismus. She was diagnosed with orbital cellulitis during her initial emergency department visit and treated with intravenous antibiotics. On her second ED visit later the same day, a diagnosis of orbital pseudotumour was made after computed tomography revealed inflammation of the sclera, optic nerve, muscle and adipose tissue within the orbit. Antibiotics were discontinued and tapering steroids were initiated, with prompt resolution of symptoms.

Key words: eye; orbital; pseudotumor; eye swelling; visual change

RÉSUMÉ

Une femme âgée de 33 ans s'est présentée au département d'urgence d'un hôpital communautaire pour des symptômes de douleur orbitale monoculaire, de photophobie et de douleur lors des mouvements de l'orbite présents depuis quatre jours. Les constatations comprenaient un chémosis, une injection conjonctivale et une restriction des mouvements de l'orbite causant un strabisme. Lors de sa visite initiale à l'urgence, on diagnostiqua chez cette patiente une cellulite orbitale qui fut traitée à l'aide d'antibiotiques intraveineux. Lors de sa seconde visite à l'urgence, un diagnostic de pseudo-tumeur orbitale fut posé après qu'un tomodensitogramme eut révélé une inflammation de la sclérotique, du nerf optique, du muscle et du tissu adipeux à l'intérieur de l'orbite. L'antibiothérapie fut interrompue et un traitement aux stéroïdes à doses dégressives fut initié, pour une résolution rapide des symptômes.

Introduction

Orbital pseudotumour is an idiopathic inflammatory process within the orbit that can pose a diagnostic challenge for emergency physicians. Since it mimics a wide array of pathologic conditions, misdiagnosis and inappropriate treatment are likely. The potential for permanent visual dysfunction makes orbital pseudotumour an ophthalmologic process that must not be overlooked. We present a case of orbital pseudotumour in a 33-year-old woman with orbital pain, photophobia and limited extraocular movement.

Case report

A 33-year-old woman with 4 days of left retro-orbital pain was referred to a community emergency department by her optometrist, who suspected orbital cellulitis. A maxillofacial computed tomography (CT) was performed. Results showed no bony abnormalities or sinus pathology, and the emergency physician administered analgesia and intravenous (IV) antibiotics. The patient was then transferred to a tertiary care centre with presumed orbital cellulitis.

On arrival to our facility, the patient complained of left-

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sided headache and sharp pain behind her left eye (OS). The pain was first noted on awakening 4 days prior and had progressed from a dull ache to a severe pain during that time. Ocular movements and intense light exacerbated the pain. The patient denied orbital foreign body sensation, trauma, fever or rash.

Her vital signs included a temperature of 37°C, pulse of 83 beats/min, respiratory rate of 16 beats/min, and blood pressure of 103/69 mm Hg. Physical examination revealed an awake, alert uncomfortable woman lying with her head under the sheets. Eye examination revealed 1+ chemosis and 2+ conjunctival injection involving the OS, but no ptosis or proptosis of either eye (OU). Pupillary exam showed that both pupils were reactive from 4 to 2 mm, but the patient experienced marked photophobia OS and severe pain with extraocular movements OS. Left extraocular movements were limited in all directions, causing strabismus during the examination. Ocular pressure was 20 mm Hg in the right eye (OD) and 23 mm Hg OS. Visual acuity was 20/25 OD and 20/30 OS with no visual field defects. Fundoscopic examination after pupillary dilation showed normal disc margins without optic nerve edema. There was no periorbital erythema, edema or cellulites, and the neck was supple with full range of motion and no adenopathy or mass. The rest of the physical examination was unremarkable.

A complete blood count was normal with a white blood cell count of $9.9 \times 10^9/L$ and no leftward shift. Erythrocyte sedimentation rate was also normal, at 11 mm/h. The maxillofacial CT showed no bony abnormalities and no fluid within the sinuses, but the orbital soft tissues were inadequately visualized. Since orbital cellulitis had not been confirmed and a space occupying lesion had not been ruled



Fig. 1. Orbital CT scan showing inflammation of the posterior sclera of the left globe, enlargement of the left optic nerve, and mild proptosis.

out, an orbital CT was performed. It showed inflammation of the posterior sclera of the left globe, enlargement of the left optic nerve and superior rectus muscle, adjacent orbital fat stranding, and left proptosis consistent with orbital pseudotumour (Fig. 1).

The ophthalmology consultant confirmed the diagnosis of orbital pseudotumour, administered an initial dose of intravenous steroids and opioid analgesics, and discharged the patient with an oral steroid taper and analgesics. A follow-up appointment in the ophthalmology clinic was arranged for the next day. Several months later, the patient reported that her symptoms resolved quickly with therapy and did not recur.

Discussion

Orbital pseudotumour is the 3rd most common disease of the orbit; only Graves' disease and lymphoproliferative diseases are more common.^{1,2} It has no racial or gender predominance but it affects primarily adults, with children representing only 6%–17% of cases.¹ Also referred to as "idiopathic orbital inflammatory syndrome" (IOIS), orbital pseudotumour is a non-granulomatous, idiopathic inflammatory process within the orbit^{3,4} that causes a variable degree of polymorphous infiltrate and fibrosis involving the orbit diffusely or targeting specific anatomic structures such as the lacrimal gland or specific muscles.^{1,2} Typical symptoms include pain, proptosis, local swelling, lid edema, conjunctival injection and erythema, but diplopia, visual loss, ptosis and extraocular dysmotility may also occur. Symptom onset is usually acute, over hours to days, but subacute (weeks) and chronic (weeks to months) presentations have also been described.¹ Often, one eye is involved, although bilateral symptoms are not uncommon.¹

Orbital pseudotumour mimics other disease processes and the differential diagnosis is broad, including orbital cellulitis, retrobulbar abscess or hematoma, sarcoidosis, tumour (primary and metastatic), Graves' disease, Wegener's granulomatosis, Sjogren's syndrome and vasculitis. Extraordinary presentations of orbital pseudotumour have been misdiagnosed as angioedema⁵ and temporomandibular joint dysfunction,⁶ the latter in a case where the inflammatory process extended through the infratemporal fossa.

Because orbital pseudotumour presents with highly variable symptoms, imaging is a critical component of diagnosis. In the emergency department, an orbital CT is the modality of choice, with magnetic resonance imaging (MRI) and B-scan ultrasonography alternative modalities. CT findings depend on which orbital structures are affected. The lacrimal gland is the most frequently affected

structure, often showing diffuse enlargement with poorly defined margins. Unfortunately, CT does not reliably differentiate lacrimal involvement from other potential causes, including bacterial inflammation.² When extraocular muscles are involved, the muscle and tendons enlarge, resulting in a tubular configuration² that contrasts with the normal tendons of thyroid ophthalmopathy. Other potential CT findings include diffuse infiltration within orbital fat, a poorly-defined orbital mass, scleral enhancement, enlargement of Tenon's space, and infiltrations along the optic nerve causing diffuse enlargement.^{2,7,8} In contrast, CT performed in cases of orbital cellulitis typically shows diffuse orbital infiltrate with decreased signal of orbital fat and may show sinus involvement, bony erosions or venous thrombosis.⁸

Corticosteroids are the mainstay of treatment, usually providing rapid regression of symptoms and decreasing the incidence of permanent disability due to sclerosis. Without treatment, the inflammation may diminish, but recurrent inflammatory episodes and residual fibrosis are common. Even with treatment, 23% to 56% of patients suffer recurrences.⁴ In cases of diagnostic uncertainty, steroid resistance or recurrence, biopsy is suggested.⁴ If the lesion is large and sclerotic, surgical debulking is an option.⁴ Other treatment options include radiation therapy and immunosuppressive agents such as cyclosporine, cyclophosphamide, azathioprine and methotrexate.⁴

Conclusion

Orbital pseudotumour is a rare ophthalmologic condition that mimics a variety of pathologic processes, creating a diagnostic dilemma for emergency physicians. A careful

history, physical examination, diagnostic suspicion and appropriate imaging are essential in arriving at the correct diagnosis.

Competing interests: None declared.

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Erratum

In our January issue we listed the names of all the reviewers who had so kindly offered their time and expertise during 2005. Regrettably, John W. King's name was missed. — The Editors