

Orbital Inflammatory Disease of the orbit

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Abstract

Purpose and method: Literature review to present the clinical course and management of orbital inflammatory disease of the orbit.

Content

Idiopathic orbital inflammation is the most commonly reported process associated with orbital inflammation. This is a localized disease process without systemic manifestations. It exists in many variations, the most common of which is known as “Idiopathic orbital inflammation (IOI)”, previously known as “Orbital pseudotumor”.

An important characteristic of all idiopathic inflammatory orbital disease is that no systemic or local cause for the inflammatory response can be identified. In this regard it is a diagnosis of exclusion. The differential diagnosis of this entity are neoplastic processes; thyroid-related orbitopathy; infectious entities such as syphilis, tuberculosis, parasitic infestations, and fungal infections; sarcoidosis; amyloidosis; and various vasculitides, including Wegener's granulomatosis and polyarteritis nodosa.

The manifestations of idiopathic orbital inflammation vary depending on which orbital structures are involved. The most common features of this process are pain and proptosis. The symptoms develop rapidly, usually within days to weeks. Generally there is chemosis and conjunctival injection often over the extraocular muscle insertions, as well as eyelid edema and ptosis. Quiet proptosis may occur but may not be found in dacryoadenitis. Diplopia and painful exacerbated during ocular motility are usually present in myositis or diffuse form of the disease, but are less common in cases of localized form. Visual loss is unusual but may be associated as well as perineuritis of the optic nerve.

The main histopathology findings are polymorphous infiltration of inflammatory cells within orbital tissue. The disease can be classified to 4 subgroups according to anatomical involvement.

1. Orbital myositis

This is characterized by the sudden onset of periorbital pain especially during ocular motility, diplopia, and conjunctival injection, usually focally located over the involved muscle. There may also be eyelid edema and proptosis. Usually single muscle is involved, but less commonly found multiple muscles involvement. The process may be recurrent despite adequate treatment. Forced-duction test is usually positive.

The most important differential diagnosis is thyroid-related orbitopathy. The extraocular muscles are involved only in the area of the muscle belly in thyroid-related orbitopathy, while the entire muscle and tendon are usually involved in idiopathic orbital inflammation. In addition, myositis is almost always painful while thyroid-related orbitopathy is usually not associated with pain.

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2. Dacryoadenitis

The affected lacrimal gland is usually tender and unilateral with enlargement that can be easily palpated in the superior lateral aspect of the orbit. This will result in an "S"-shaped deformity of the upper eyelid. Proptosis with inferomedial globe displacement can be found. Bilateral involvement suggests the presence of a systemic disease such as sarcoidosis, lymphoma or Sjögren's syndrome.

3. Anterior orbital inflammation

This entity is characterized by acute or subacute inflammation of the globe, in particular the sclera, Tenon's capsule and surrounding orbital tissue. It is usually associated with decreased vision either from uveitis with or without associated exudative retinal detachment or papillitis and perineuritis. Chemosis, conjunctival injection (especially episcleral vessels), eyelid edema can also be found in this process. Rarely there may be anterior segment cell and flare and even a sterile hypopyon.

4. Apical orbital inflammation

The inflammation involving the orbital apex is associated with marked restriction of ocular motility as well as pain and minimal proptosis. Boring and persistent pain behind the globe is characteristic. There are often minimal signs of inflammation such as eyelid edema, chemosis, or conjunctival injection. Decreased vision and an afferent pupillary defect are frequently either from perineuritis or compressive neuropathy due to soft tissue edema. Computed tomography (CT) of the orbit will demonstrate a diffuse inflammatory process in the posterior orbit with extension into the adjacent orbital fat. The inflammation may be noted to extend forward along the optic nerve or posteriorly through the superior orbital fissure.

In general, idiopathic inflammation of the orbit is well response to corticosteroids. Prednisone in an oral dose of 80 to 100 mg will usually result in a marked improvement within 2 to 3 days. The treatment should be continued for 2 to 3 weeks, and then slowly tapered over an additional 3 weeks. Failure to do this will result in recurrence of the orbital inflammation.

Although in the vast majority of cases corticosteroids are curative, a small number of patients will either have recurrence of their disease process or fail to adequately respond to this treatment. In the recurrence group, a second course of corticosteroids can be instituted over a longer period of time and with a prolonged taper. In the failure group, immunosuppressive therapy with cyclophosphamide or radiotherapy can be considered. The greater the amount of fibrosis that is present within the orbit, the less responsive the inflammatory process is to steroid administration. It is important for tissue biopsy in patients that fail to respond to an adequate course of corticosteroids.