

Pterygomaxillary extension of orbital pseudotumor. Case report

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SUMMARY: Pterygomaxillary extension of orbital pseudotumor. Case report.

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Idiopathic pseudotumor (IP) is a non-specific, non-neoplastic, inflammatory process without identifiable local or systemic causes. IP most commonly affects the orbit, comprising 5% to 8% of all orbital masses with occasional extension to other areas of the skull base. The ophthalmology literature has detailed reviews of IP as a space-occupying intraorbital lesion, but lesion involving only the skull base and sparing the orbit is exceptional. Understanding the presentation, work-up, and characteristics of the lesion is important for proper diagnosis and treatment.

We report a rare case of orbital IP with extension into infratemporal fossa. Clinical and radiological findings and surgical approach are discussed.

RIASSUNTO: Estensione nello spazio pterigo-mascellare di pseudotumore orbitario. Caso clinico.

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Lo pseudotumore orbitario è un'infiammazione benigna, aspecifica e idiomatica, localizzata nell'orbita e con eccezionale estensione alla base cranica. La sua eziopatogenesi non è conosciuta, definirne la natura istologica è importante per un corretto trattamento.

Riportiamo un raro caso di pseudotumore orbitario con estensione nella fossa infratemporale discutendone gli aspetti clinici e terapeutici.

KEY WORDS: Orbit - Pseudotumor - Surgery.
Orbita - Pseudotumor - Chirurgia.

Introduction

Pseudotumor of the orbit is a clinical condition due to idiopathic inflammation of the orbital contents not related to specific infection, or neoplasm, or systemic disease. The extension of the lesion beyond the orbit or even intracranially is quite rare. Such an extension consists of a non-specific inflammatory tissue of the orbit that extends through one or more foramina into the adjacent extraorbital or intracranial structures. The middle cranial fossa and cavernous sinus are the two mo-

st common locations into which extensions are observed (1). A direct intracranial spread can occur through the superior orbital fissure, the inferior orbital fissure or the optic canal. An extension of the lesion into the pterygopalatine and infratemporal fossa probably reflects a chronic inflammatory lesion extending through the inferior orbital fissure (2). When a pseudotumor invades the cavernous sinus, the process is usually considered to be part of Tolosa-Hunt syndrome (3).

Orbital IP is a disease of all age groups and both sexes. Unilaterality is the rule except in children, where synchronous bilateral disease is present. Symptoms typically include pain, photophobia, proptosis, eyelid swelling, chemosis and diplopia. Although considered a benign, self-limited process, it may endanger the optic nerve leading to vision loss. Presentation of nonorbital skull base IP varies by location but can include facial asymmetry, facial pain, nasal obstruction, dyspnea, or dysphagia.

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Case report

A 72-year-old female was admitted to our hospital with right proptosis, facial pain and intermittent headache for 4 months. Physical examination did not reveal any abnormality and blood routine investigations were normal. She was fully conscious alert and oriented without motor system deficits.

A CT of the orbit revealed a soft tissue orbital mass with homogeneous mild enhancement with bone erosion and extension into the adjacent infratemporal fossa and pterygopalatine space (Fig. 1). A lateral orbitotomy was performed showing a fibrous lesion that was completely removed (Fig. 2) with uneventful postoperative period; histopathology analysis confirmed the diagnosis of orbital pseudotumor revealing a chronic inflammatory cell infiltration with plasma cells, lymphocytes and diffuse area of calcification and fibro connective tissue.

High dose corticosteroids (dexamethasone 16 mg/die) were given with complete resolution of symptoms and radiographic abnormalities at 3 month follow-up.

Discussion and conclusion

The term orbital pseudotumor originally included all nonneoplastic disorders, but is now limited to idiopathic lesions. The incidence of orbital pseudotumors has been reported to be 8% of all orbital mass lesions (4). The cause of orbital IP remains a mystery, and three theories currently exist. First, and most likely, is an autoimmune reaction mediated by both B and T lymphocytes (5) and these reactions have been linked to viral infections, sinus infections, and mollicute-like organisms (cell wall deficient bacteria); some authors have implicated viral infections with Epstein-Barr virus (EBV) or human herpes virus-8 as playing a role in pathogenesis (6). The second theory is the classic thought that orbital IP is an infectious process as a result of sinusitis or syphilis. The onset of disease has been associated with upper respiratory infection (URI) or other viral illness; however, no definitive organism has been associated reliably with orbital IP. The third theory is based on pathological findings of fibroproliferative disorders; these disorders involve aberrant production of fibrogenic cytokines and characteristic histological findings include mixed inflammatory infiltrates with polyclonal lymphocytes, macrophages, polymorphonuclear cells, plasma cells, eosinophils, occasional histiocytes, and a varying bed of fibrosis.

The radiological findings in a pseudotumor are characterized by inflammatory changes in the various intraorbital structures, such as the globe, the lacrimal

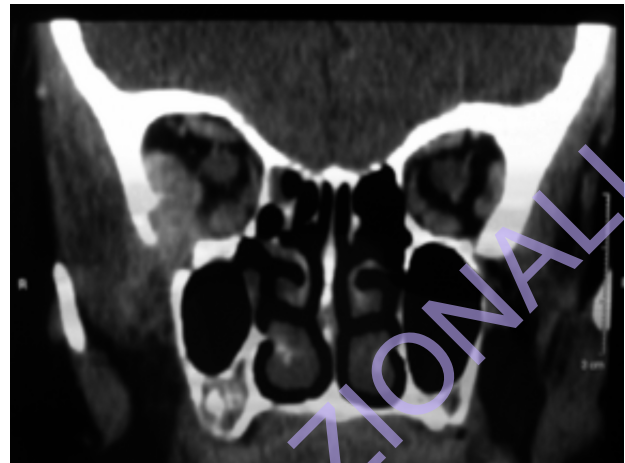


Fig. 1 - Coronal CT shows a right orbital mass with extension into pterygopalatine space with bone erosion.

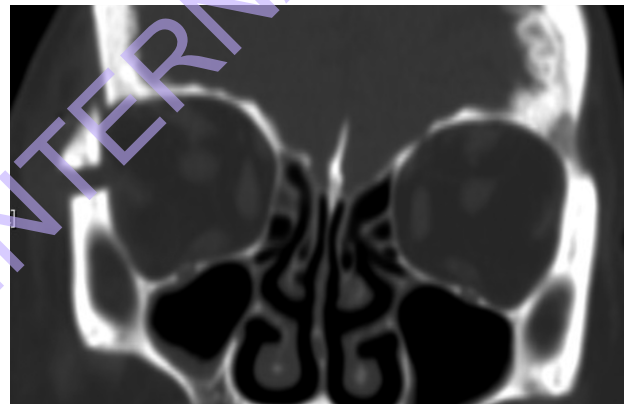


Fig. 2 - Postoperative CT scan demonstrating no residual mass.

glands, the extraocular muscles, the orbital fat and the optic nerve. The CT findings do not allow idiopathic orbital inflammation to be unequivocally distinguished from other orbital mass lesions. MRI shows a hypointense lesion on the T1- and T2-weighted images and shows marked gadolinium enhancement. Fat suppression techniques clearly show an optic nerve sheath lesion and intraorbital inflammation (7). Differential diagnosis should include lymphoma, plasmocytoma, and advanced sinus malignant tumors (melanoma, sarcomas, epidermoid carcinoma).

Finally, we think that high dose corticosteroids always have to follow the biopsy for a period than at least three months.

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