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Intracranial Extension of an Idiopathic Orbital Inflammatory Pseudotumor

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Summary: Idiopathic orbital inflammatory pseudotumor (IOIP) with endocranial extension is very unusual. The authors used CT and MR to diagnose IOIP and demonstrate the presence of intracranial extension of orbital and lacrimal gland lesions. While providing additional evidence of IOIP having intracranial extension, this case report emphasizes the need to include IOIP as a possible differential diagnosis when radiologic explorations reveal lesions extending from the orbit to intracranial structures.

Index terms: Orbits, magnetic resonance; Orbits, neoplasms

Idiopathic orbital inflammatory pseudotumor (IOIP) is a lesion of unknown etiology involving focal or multifocal intraorbital structures (1), and lacrimal gland inflammation (dacryoadenitis), which is relatively frequent (2); however, extension of the lesion beyond the orbit or intracranially is very rare (3–5). Usually, a small extension is localized around the superior orbital fissure. The IOIP appears as an intraorbital mass responsible for a severe proptosis at clinical presentation. Dramatic response to steroids and biopsy results suggest the precise diagnosis. Typically, the pathology reveals a heterotopic cellular component with a mixture of small lymphocytes, plasma cells, and histiocytes. In addition, true germinal centers, vasculitis, granulomas, and fibrosis are also frequently present (6). Systemic disease like Wegener granulomatosis, polyarteritis nodosa, or multifocal fibrosclerosis (7) may have the same clinical picture with the diagnosis made on patient follow-up.

Previous studies based on computed tomography (CT) techniques have reported the involvement of both intraorbital and intracranial structures in IOIP (4, 5). In these reports, the orbital lesions described were apical, bilateral, or superomedial (5). The present report describes a case of unilateral pseudotumor involving the orbit, including the lacrimal gland, plus an important

endocranial extension. To our knowledge, the latter feature has not been previously reported. The combined use of magnetic resonance (MR) and CT techniques, in our case, has been very useful in precisely evaluating the extent of the lesions.

Case Report

A 23-year-old man referred to our department in January 1990 had a history of episodes of asthenia, diplopia, and progressive left proptosis which started in February 1989. Physical examination at that time showed a complete ophthalmoplegia, and visual acuity was 20/20 with normal fundoscopy. CT examination revealed an enhancing lesion in the outer superior quadrant of the left orbit with an endocranial component. Treatment with steroids (prednisone, 40 mg/day) was initiated and there was complete resolution of the ophthalmoplegia. In October 1989, examination revealed proptosis of the left eye with chemosis and corneal ulcerations. Visual acuity was 1/20 and papillary edema was found at fundoscopy. CT and MR showed an endocranial component of the lesion extending along the lateral wall of the left cavernous sinus. Cerebral angiography was normal and left orbital phlebography showed inferomedial displacement of the superior ophthalmic vein. The patient was then referred to our department.

Physical examination revealed asthenia and thyroid enlargement. There was left facial pain, proptosis with corneal lesions, ophthalmoplegia, and abolition of thyrotropin pupillary reaction. Thyroid hormones (T3, T4) and thyrotropin levels were normal.

Thyroid scanning and ultrasonography showed an enlarged gland without focal abnormalities. CT demonstrated an enlarged, homogeneously enhancing left lacrimal gland contiguous with a superolateral extraconal lesion extending through the superior orbital fissure endocranially and along the left frontotemporal dura mater and the lateral wall of the cavernous sinus (Fig. 1A). There was also extension to the pterygopalatine fossa, probably through the inferior orbital fissure, and lateral extension to the infratemporal

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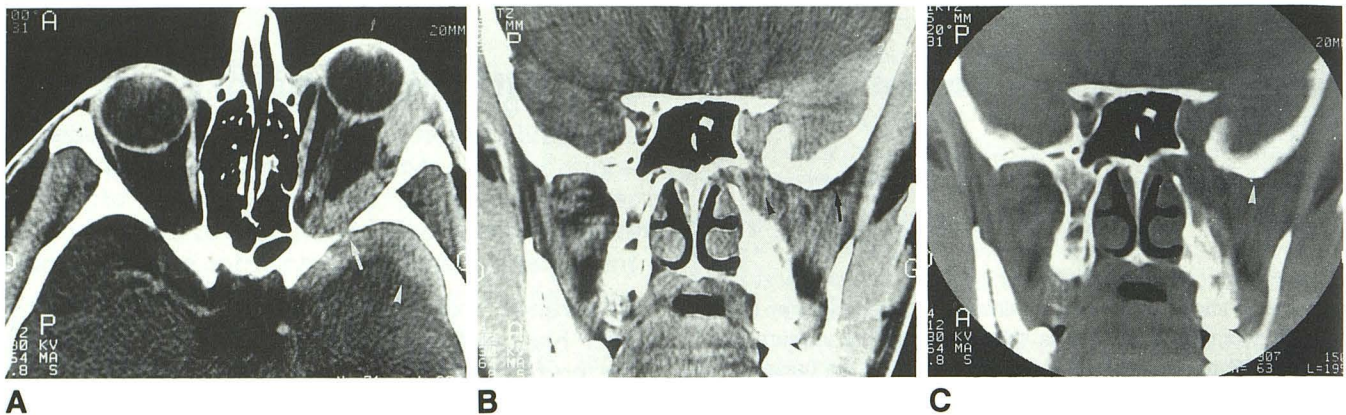


Fig. 1. Axial and coronal CT after contrast injection. In A, the orbital and endocranial component (arrowhead) of the lesion are in continuity at the level of an enlarged superior orbital fissure (arrow). In B, The lesion extends into the infratemporal fossa (arrowhead) through the inferior orbital fissure (arrow). There is marked hyperostosis of the sphenoid bone as shown in C (arrowhead).

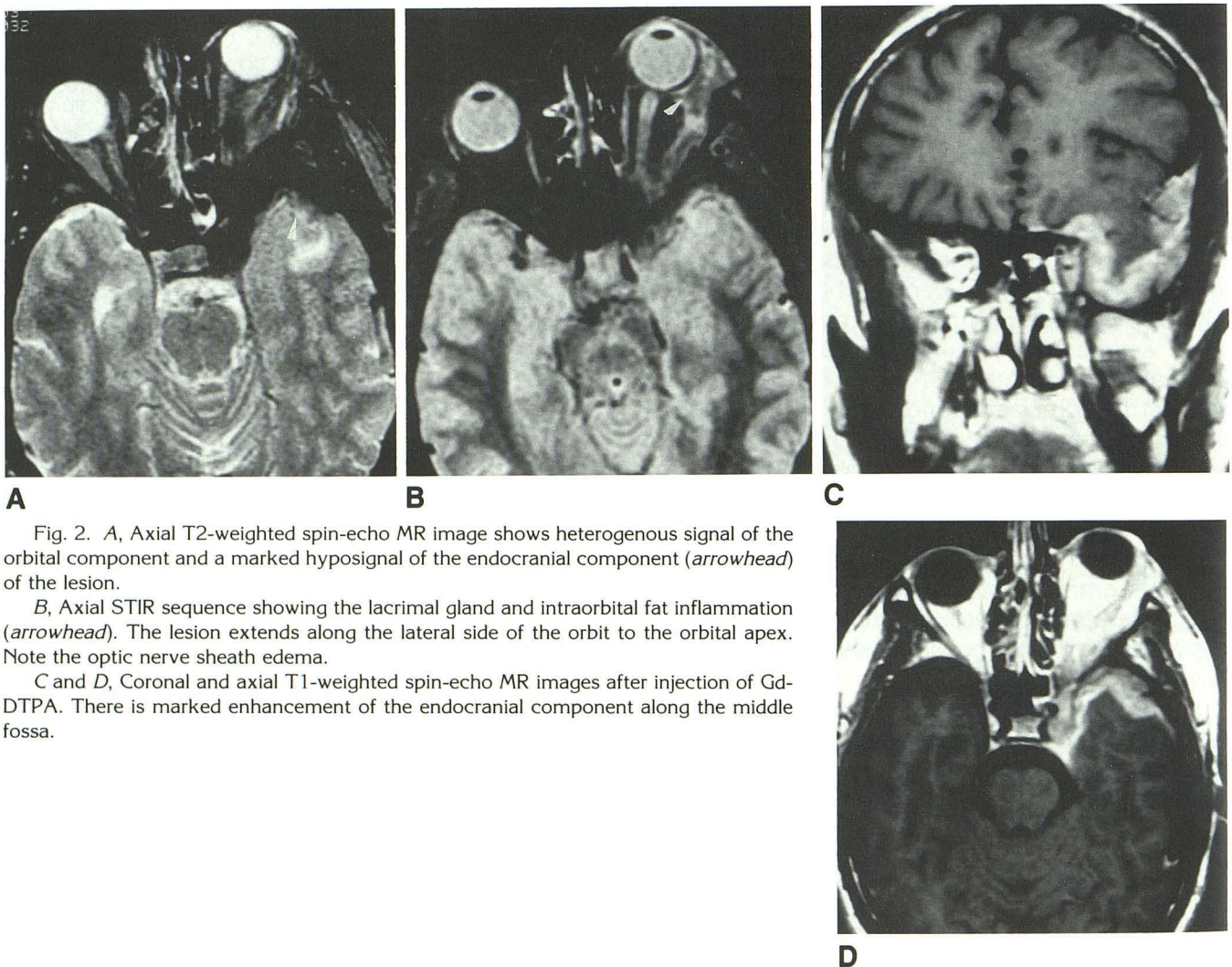


Fig. 2. A, Axial T2-weighted spin-echo MR image shows heterogenous signal of the orbital component and a marked hyposignal of the endocranial component (arrowhead) of the lesion.

B, Axial STIR sequence showing the lacrimal gland and intraorbital fat inflammation (arrowhead). The lesion extends along the lateral side of the orbit to the orbital apex. Note the optic nerve sheath edema.

C and D, Coronal and axial T1-weighted spin-echo MR images after injection of Gd-DTPA. There is marked enhancement of the endocranial component along the middle fossa.

fossa (Fig. 1B). The sphenoid bone showed signs of sclerosis and remodeling (Fig. 1C).

Cranial MR was performed on a 1.5-T system using a standard head coil. Sagittal spin-echo T1-weighted, axial

proton density, and T2-weighted and short T1 inversion recovery (STIR) sequences were obtained (Figs. 2A and 2B). Additional post-Gd-DTPA spin-echo sagittal, coronal and axial T1-weighted images were obtained (Figs. 2C and

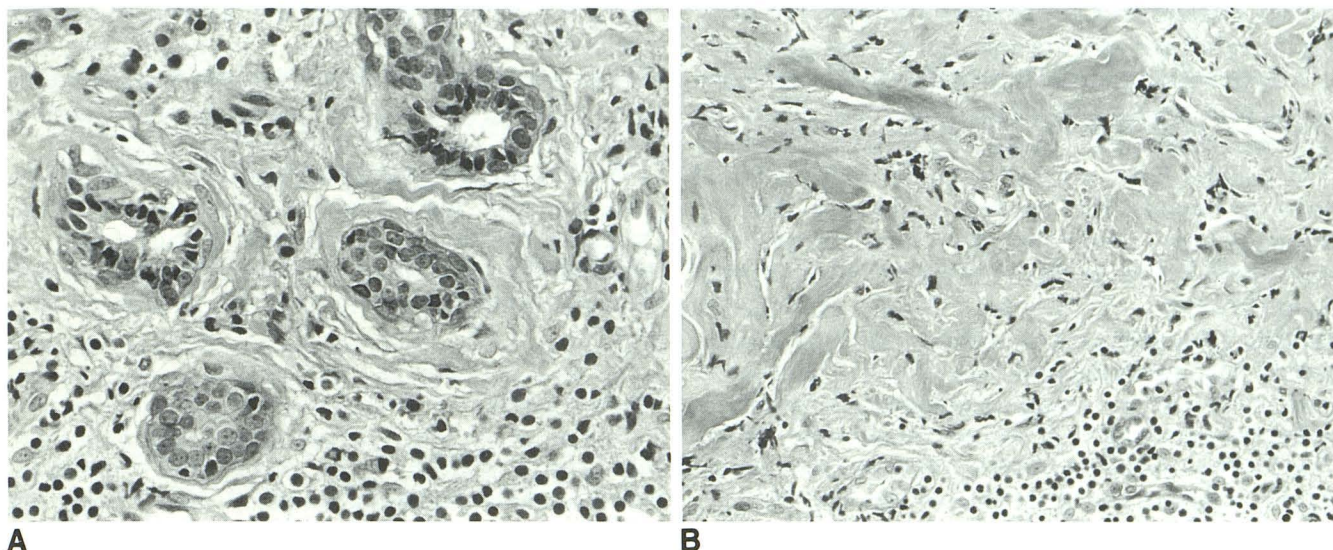


Fig. 3. Masson stain, trichrome X250.

A, Lacrimal gland acini surrounded by mononuclear cells described as lymphocytes and macrophages embedded in a fibrous stroma.
B, Chronic inflammatory infiltrate associated with fibrocollagenous tissue.

2D). The lesion appeared hypointense on T1- and T2-weighted sequences and showed marked enhancement on post-Gd-DTPA T1-weighted sequences. The STIR sequences showed a heterogeneous hypersignal of the intraorbital lesion.

A left superolateral orbitotomy was performed. A hard and infiltrated grayish-white lacrimal gland with surrounding tissue was removed and pathology revealed a fibrocollagenous stroma with mononuclear cells surrounding lacrimal gland acini (Figs. 3A and 3B), thereby confirming the diagnosis of IOIP of the lacrimal gland. The patient was given high doses of steroid (Cortancyl, 60 mg/day), and there was dramatic reduction of the proptosis and ophthalmoplegia. He was discharged on long-term steroid therapy. The patient went back to his native country. One month later, he was disease-free, but we have no long-term follow-up data.

Discussion

Idiopathic orbital inflammatory pseudotumor is an inflammatory lesion of the orbit without any recognizable local or systemic causes (2). Clinically, the patient exhibits acute onset of proptosis, chemosis, pain, diplopia, and impaired ocular motility or, in some cases, a subacute or chronic onset of these symptoms and signs. Many forms have been described depending on the structures involved: myositis, dacryoadenitis, periscleritis, trocheitis, perineuritis, or diffuse (2).

A good correlation between clinical signs and location of the lesions has previously been reported by Nugent (7), who described five patterns of anatomical involvement: anterior, posterior,

diffuse, lacrimal, and myositic, with the lacrimal lesion being the most frequent. Characteristically, the onset of symptoms and signs is acute and the lesion is very sensitive to high doses of prednisone, but there may be recurrences, and chronicity can develop.

Acute forms are usually very sensitive to high doses of steroid, but in sclerosing forms, the treatment may be more difficult. Radiotherapy may be indicated when corticosteroids fail or are contraindicated (8), and may give good results when biopsy reveals cellular rather than fibrotic predominance (9). Surgery may be indicated when other approaches have failed (2). Lesions are commonly restricted to the orbit. However, extension beyond the orbit can occur in cases of inexorable sclerosing orbital inflammation. When these lesions are restricted to the lacrimal gland, they are usually characterized by a swollen oblong gland with normal contiguous bone. If endocranial extension is present, orbital lesions are either restricted to the apex of the orbit or are extensive and bilateral with paranasal involvement (3–5). However the present report indicates that even lesions involving more anterior structures, such as the lacrimal gland, can be associated with intracranial extension. In this case, a precise assessment of the extent of orbital and endocranial lesions using both MR and CT was valuable in differentiating IOIP from other similar clinical entities.

In its acute form, idiopathic orbital inflammatory pseudotumor is easily distinguishable from

Grave disease and lymphoid tumors of the orbit (2). In subacute and chronic forms, the diagnosis is more difficult and a biopsy may be necessary to rule out a neoplasm. The differential diagnosis includes malignant lesions of the lacrimal gland that usually have a short evolution with a tendency to invade the muscle cone and destroy adjacent bone. They can show hyperostosis and thickening of the involved bone in cases of mucoepidermoid carcinomas (10). Sometimes chronic idiopathic orbital inflammatory pseudotumor can simulate lymphoma and a biopsy is necessary for the diagnosis, particularly when there is no history of acute onset. MR signal intensity of idiopathic orbital inflammatory pseudotumor and metastasis have been reported to be different; thus, MR imaging may add specificity to the diagnosis (11).

In the later stage of Grave disease, particularly when advanced extraocular muscle swelling is present, lacrimal gland enlargement is also seen (2, 12), but the tendons of the extraocular muscles are usually spared and the intraorbital fat is not inflamed (2). Furthermore, intracranial extension is not seen. Hypertrophic tuberculosis pachymeningitis may also involve the orbital apex (13), but involvement of the lacrimal gland is rare and seen only with extensive lesions invading the orbit. In extremely aggressive idiopathic orbital inflammatory syndrome, the pathologic diagnosis should be reevaluated to exclude a malignant fibrous inflammatory histiocytoma (10).

Bone lesions are particularly well defined by CT but MR is superior in delineating the soft-tissue lesions in the orbit and their endocranial extension. In our case, the lesion generated low signal in T1- and T2-weighted sequences, probably reflecting fibrotic changes (1, 11), and enhanced significantly after contrast injection. Fat suppression techniques (14) showed clearly the optic nerve sheath edema and the intraorbital inflammation. Bony changes reflected by hyperostosis and remodeling favored a long-standing

benign process, whereas the extension of the lesion to the middle cranial, pterygopalatine, and infratemporal fossae probably reflected an extensive and chronic inflammatory lesion extending through the various foramina of the orbit (15).

In conclusion, we have reported a case of an unusual IOIP with major intracranial extension, stressed the uniqueness of this feature, and discussed the differential diagnosis of other orbital lesions more commonly extending intracranially.

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