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Case Report: Idiopathic Orbital Inflammatory Pseudotumor (IOIP)

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Abstract **Case Report**

Idiopathic Orbital Inflammatory Pseudotumor (IOIP) presents a significant diagnostic challenge in neuroradiology, often mimicking orbital malignancies. We report the case of a 23-year-old female with progressive unilateral exophthalmos. This report highlights the critical role of multi-modal imaging specifically MRI in characterizing inflammatory lesions and guiding clinical management to avoid unnecessary invasive procedures.

Keywords: Idiopathic Orbital Inflammatory Pseudotumor, IOIP, MRI, proptosis, differential diagnosis, orbital myositis.

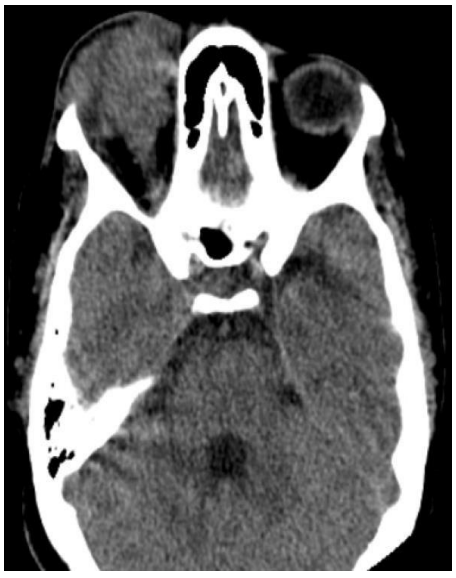
INTRODUCTION

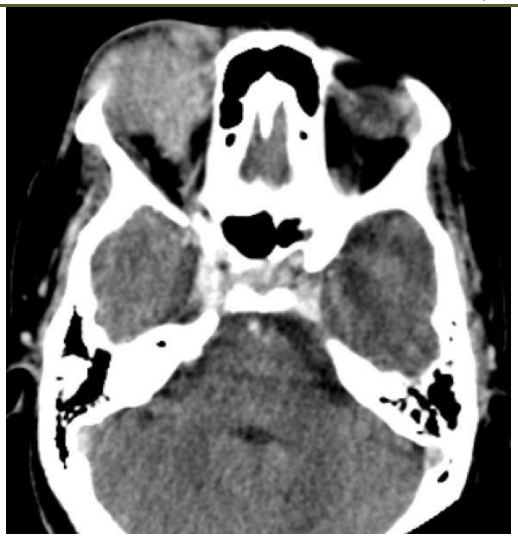
Orbital Inflammatory Pseudotumor (IOIP), or Idiopathic Orbital Inflammatory Disease (IOID), is a non-neoplastic, non-granulomatous inflammatory lesion of the orbit. As a diagnosis of exclusion, IOIP requires careful differentiation from orbital neoplasms, infections, and systemic inflammatory diseases such as IgG4-related ophthalmic disease. The radiological approach is paramount to steering clinicians away from invasive surgical interventions toward effective medical management.

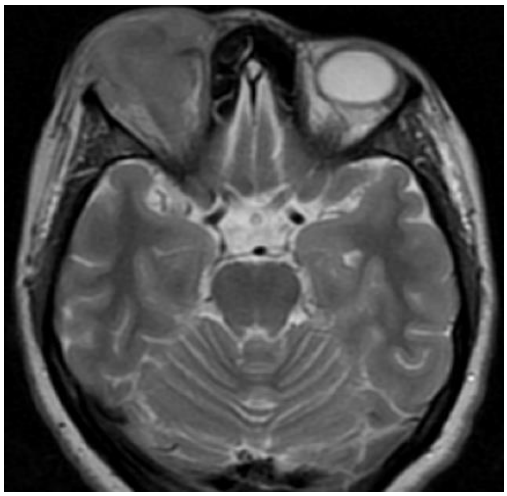
CASE PRESENTATION

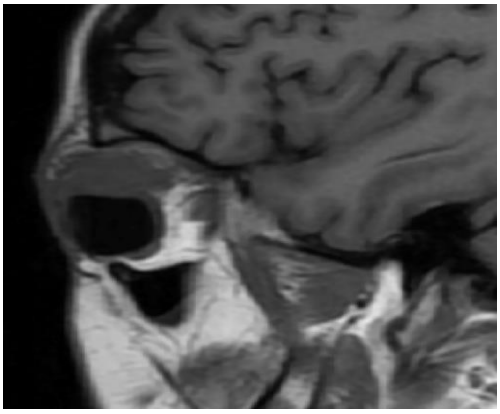
A 23-year-old female with a known history of hypothyroidism (non-compliant with medication) presented with progressive right-sided unilateral proptosis evolving over six weeks. Physical examination revealed clear proptosis without signs of orbital cellulitis (no fever, normal white blood cell count). Visual acuity and ocular motility remained intact. Surgical biopsy was performed following imaging, which confirmed non-specific chronic and acute inflammation.

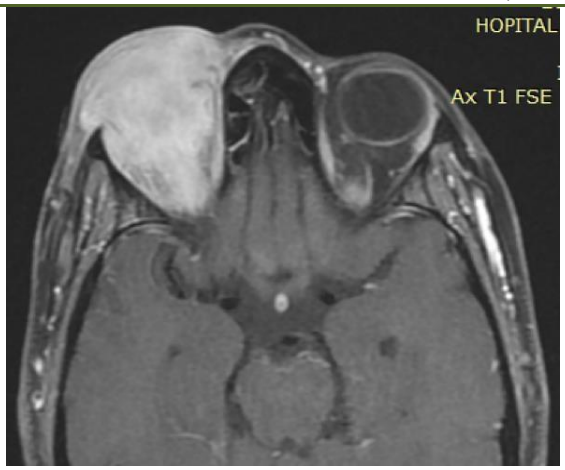
Clinical Images:











Radiological Description

Computed Tomography (CT)

CT was the initial modality, revealing a soft-tissue density mass localized to the superior orbit. The mass showed well-defined margins while demonstrating infiltrative behavior toward the surrounding fat. Crucially, there was no bone destruction or extension into intracranial spaces, findings that strongly argue against malignant primary orbital tumors or invasive fungal infections.

Magnetic Resonance Imaging (MRI)

MRI provided detailed characterization of the lesion:

- **Signal Intensity (T1-weighted):** Iso-intense signal compared to extra-ocular muscles, allowing for clear demarcation of the lesion's boundaries.
- **Signal Intensity (T2-weighted):** Heterogeneous intermediate signal intensity. This heterogeneity reflects the presence of fibrotic components intermingled with areas of active inflammation.
- **Post-Contrast Dynamics:** Intense and homogeneous enhancement was observed following gadolinium administration. This intense uptake is indicative of high vascularity within the reactive granulation tissue.
- **Anatomic Involvement:** The superior rectus muscle appeared thickened and incorporated into the inflammatory mass, suggestive of myositis involvement. The mass induced significant compression and deformation of the ocular globe, explaining the clinical proptosis.

Differential Diagnosis

Pathology	Key Distinguishing Features
Orbital Lymphoma	Typically homogeneous; restricted diffusion; usually older patient demographic.
IgG4-Related Disease	Often bilateral; systemic involvement; requires specific serum IgG4 testing.
Sarcoidosis	Requires histologic evidence of non-caseating granulomas.
Granulomatosis with Polyangiitis	Associated with sinus destruction and positive serology (ANCA).

Management and Evolution

The diagnosis was established by excluding malignant, infectious, and specific systemic inflammatory etiologies. Management consisted of high-dose intravenous corticosteroid bolus therapy followed by a gradual oral taper. The patient demonstrated an excellent therapeutic response, characterized by rapid resolution of proptosis and orbital discomfort within days.

CONCLUSION

IOIP requires high clinical suspicion and a systematic radiological approach. MRI remains the imaging standard, providing crucial information on enhancement patterns and anatomic involvement that guides the medical, rather than surgical, management of this condition.

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