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SEEING THE UNSEEN: A DIAGNOSTIC JOURNEY INTO ORBITAL LYMPHOMA

Radio-Diagnosis

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ABSTRACT

Introduction: Primary orbital lymphoma (POL) is a rare, extranodal malignancy arising within the orbit, accounting for approximately 55% of orbital malignancies. It predominantly consists of mucosa-associated lymphoid tissue (MALT) lymphoma but may also include other subtypes like diffuse large B-cell lymphoma (DLBCL). POL typically presents in older adults with painless, progressive proptosis and can be diagnosed using imaging modalities such as CT, MRI, ultrasound, and PET-CT. **Case Presentation:** A 72-year-old male presented with a one-month history of non-pulsatile proptosis of the left eye and blurring of vision. There was no history of trauma or prior malignancy. MRI revealed a heterogeneously enhancing extraconal lesion causing lateral displacement of the globe, with extension into the nasal cavity, ethmoidal air cells, and frontal sinus. Differential diagnoses included lymphoma, nerve sheath tumor, rhabdomyosarcoma, and metastases. Biopsy confirmed DLBCL, and systemic workup ruled out secondary lymphoma. The patient was diagnosed with primary intraorbital lymphoma and initiated on chemotherapy with Rituximab, Doxorubicin, Vincristine, and Cyclophosphamide. **Discussion:** POL commonly presents as a painless orbital mass, with imaging features of a well-defined, homogenous lesion without bony destruction. It is crucial to differentiate it from orbital pseudotumor, metastatic tumors, and lacrimal gland neoplasms. MRI findings include iso- to hypointensity on T1, hyperintensity on T2, and moderate contrast enhancement, while PET-CT aids in staging. Early diagnosis through imaging and biopsy is essential for prompt management. **Conclusion:** Treatment of POL depends on disease extent, with localized cases managed by radiotherapy and systemic involvement requiring chemotherapy and immunotherapy. The prognosis is favorable for MALT lymphoma, while high-grade variants like DLBCL necessitate aggressive treatment. Early detection and tailored management strategies are vital for optimal patient outcomes.

KEYWORDS

INTRODUCTION:

Primary orbital lymphoma (POL) is a rare, extranodal, low-grade malignant tumor arising within the orbit. It accounts for approximately 55% of orbital malignancies and is the most common orbital tumor in adults over 60 years of age (1). It predominantly consists of mucosa-associated lymphoid tissue (MALT) lymphoma, though other subtypes, such as diffuse large B-cell lymphoma (DLBCL), can also occur (2).

Imaging Modalities for Diagnosis

1. Computed Tomography (CT)

Findings: Well-defined, homogenous, iso- to hyperattenuating soft-tissue mass (3).
Common Locations: Superior-lateral orbit, lacrimal gland, or extraconal regions (4).
Enhancement: Mild to moderate enhancement with contrast.
Bone Involvement: Typically does not cause bony destruction but may mold around adjacent structures (5).

2. Magnetic Resonance Imaging (MRI)

- **T1-weighted**: Iso- to hypointense relative to muscle (6).
- **T2-weighted**: Hyperintense, reflecting a soft tissue mass (6).
- **Post-contrast**: Moderate homogeneous enhancement.
- **DWI (Diffusion-Weighted Imaging)**: Restricted diffusion, aiding differentiation from benign lesions (7).

3. Ultrasound (US)

Findings: Hypoechoic, well-defined soft-tissue mass with mild internal vascularity on Doppler imaging (8).

4. Positron Emission Tomography (PET-CT)

Findings: Increased FDG uptake, useful for staging and systemic involvement (9).

Key Radiological Differentiation

- **From Orbital Pseudotumor**: POL lacks pain, does not respond to steroids, and has a well-defined, homogenous mass (10).
- **From Metastatic Tumors**: POL is typically localized without widespread metastasis at presentation (11).
- **From Lacrimal Gland Tumors**: POL molds around structures, while lacrimal gland tumors cause displacement and infiltration (12).

CASE REPORT:

A 72 year old male presented with the complaints of non pulsatile proptosis of left eye for the last 1 month with blurring of vision
No history of trauma to eye

Medical history: Not a known case of malignancy

Surgical history: No previous major surgeries

Habits: Tobacco chewer for last 50 years

Comorbidities: Known hypertensive (On treatment) and non diabetic

The patient was scanned using 3T MRI Machine

Brain was studied in Axial, Sagittal and Coronal planes with 5 mm contiguous slices.

Sequences: Spin Echo T1, Turbo spin echo T2 and FLAIR.

10 ml of gadolinium I.V. contrast was used.

MRI FINDINGS:

Heterogeneously enhancing T2 and FLAIR hypointense, T1 isointense lesion showing diffusion restriction showing involving extraconal compartment of left orbit medially causing mass effect on globe resulting in its lateral displacement.

Medially, the lesion shows erosion of medial wall of orbit with extension into nasal cavity resulting in erosion of superior and middle turbinates involving few of ethmoidal air cells on right side with superior extension into frontal sinus.

On the basis of imaging findings, following differentials were considered:

From extraconal compartment with extension into paranasal sinuses

- Lymphoma
- Nerve sheath tumour
- Rhabdomyosarcoma
- Intraorbital ewings sarcoma
- Metastases
- Olfactory neuroblastoma

Less likely tumours from paranasal sinuses with extension into extraorbital compartment - Sinonasal squamous call carcinoma, sinonasal adenocarcinoma, Sinonasal undifferentiated carcinoma, sinonasal mucosal melanoma

Biopsy of the lesion: Diffuse Large B Cell Lymphoma
Patient was advised for CECT thorax and abdomen to rule out primary vs secondary lymphoma which were normal

Patient was finally diagnosed as primary intraorbital lymphoma and started on chemotherapy with Rituxumab, Doxorubicin, Vincristine and Cyclophosphamide

DISCUSSION:

Primary intraorbital lymphoma (PIOL) is a rare, extranodal malignancy that arises within the orbit without systemic involvement at diagnosis, most commonly presenting as MALT lymphoma (1,3). It typically affects older adults and manifests as a painless, slowly progressive proptosis, often accompanied by diplopia, periorbital swelling, or ptosis, but without systemic B symptoms (4,5). Radiologically, CT scans show a well-defined, homogenous soft tissue mass without bone destruction, while MRI reveals iso- to hypointense signals on T1, hyperintensity on T2, and moderate contrast enhancement (6,7). Ultrasound demonstrates a hypoechoic, well-circumscribed lesion with mild vascularity, and PET-CT may indicate increased FDG uptake, useful for detecting systemic spread (8,9). The primary differential diagnoses include orbital pseudotumor, which presents with pain and steroid responsiveness, metastatic tumors, which are more aggressive and infiltrative, and lacrimal gland tumors, which cause distinct globe displacement (10-12).

TREATMENT OPTIONS:

Treatment depends on disease extent, with radiotherapy being the mainstay for localized cases, while chemotherapy and immunotherapy (such as Rituximab) are used for high-grade or disseminated disease (13). Prognosis is generally favorable for MALT lymphoma, but more aggressive subtypes require intensive treatment, emphasizing the importance of early imaging, biopsy confirmation, and tailored management strategies for optimal patient outcomes (14,15).

CONCLUSION:

Primary intraorbital lymphoma is a rare tumor with different multiple patterns of involvement. Radiologists must be aware of the different imaging scenarios to optimize imaging protocols to support early and accurate diagnosis, besides helping in the treatment and follow-up of the patients (16)

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CONFLICTS OF INTEREST:

There are no conflicts of interest.

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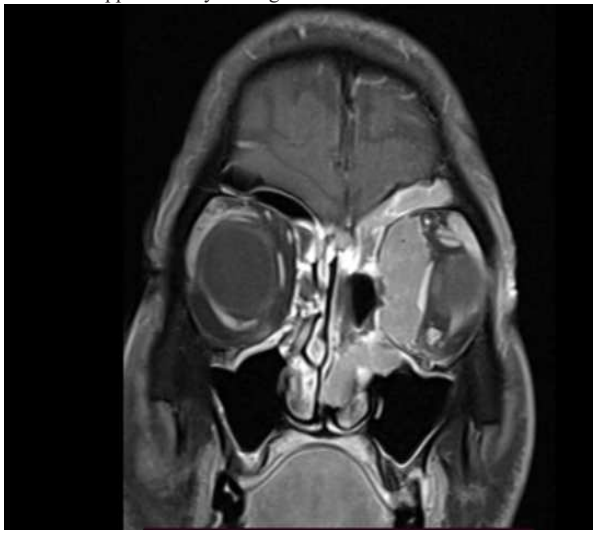

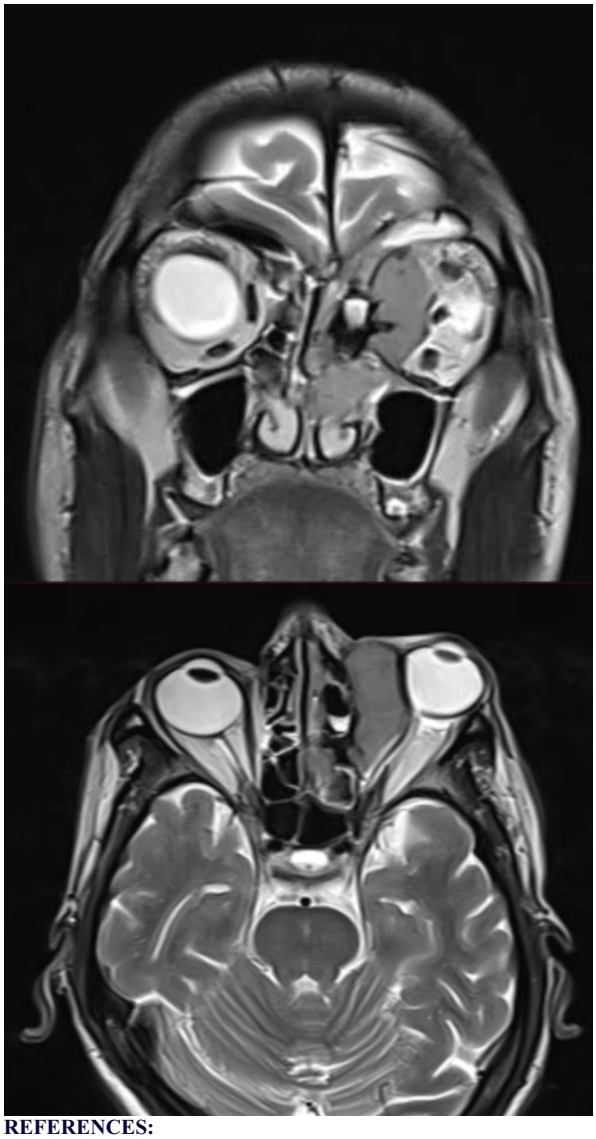

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