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A HIDDEN MENACE BEHIND A PROPTOSED EYE: A CASE REPORT OF SMALL BLUE CELL CARCINOMA-ADAMANTINOMA LIKE EWING'S SARCOMA CAUSING PROPTOSIS IN AN ADULT.

Ophthalomology

Dr. Vinutha Shree R Post Graduate, Department of Ophthalmology, Rio, Minto Ophthalmic Hospital, BMCRI, New Tharagupete, Bangalore, Karnataka-560002.

Dr. Chinmayee Thrishulamurthy Professor, Department of Ophthalmology, Rio, Minto Ophthalmic Hospital, BMCRI, New Tharagupete, Bangalore, Karnataka-560002.

ABSTRACT

Background: Proptosis can result from a wide range of orbital pathologies, including inflammatory, vascular, and neoplastic lesions. Among these, small blue cell carcinoma–adamantinoma-like Ewing's sarcoma (AL-EWS) occurring in an adult is an extremely rare and aggressive tumour that can mimic benign orbital conditions, leading to delayed diagnosis and management. **Objective:** To report a rare case of AL-EWS presenting as progressive proptosis of the left eye and discuss its clinical, radiological, and histopathological features, along with management and outcomes. **Methods:** A case of a 42-year-old female presenting with swelling and outward protrusion of the left lower eyelid for 2 months was evaluated at a tertiary ophthalmology centre. Detailed clinical examination, imaging studies (CT/MRI), and histopathological analysis including immunohistochemistry were performed to establish the diagnosis. Management strategies and follow-up outcomes were documented. **Results:** The patient presented with gradual proptosis, diminution of vision, and pricking sensation, with restriction of ocular movements but no redness, discharge, diplopia, or history of trauma. Imaging revealed a [describe location/extent of orbital mass]. Histopathology showed sheets of small round blue cells with adamantinoma-like features, and immunohistochemistry was positive for CD99, cytokeratin, and FLI-1, confirming AL-EWS. Surgical intervention was performed, and [outcome – e.g., visual improvement or follow-up status] was noted. **Conclusion:** AL-EWS should be considered in the differential diagnosis of progressive proptosis, especially in adults with no history of trauma. Early recognition and prompt multidisciplinary management are critical due to the tumour's aggressive nature and potential for local invasion and systemic spread.

KEYWORDS

Proptosis, Orbit, Small Blue Cell Tumour, Adamantinoma-like Ewing's Sarcoma, Case Report, Orbital Tumour

INTRODUCTION

Proptosis is a common manifestation of orbital pathology and may result from inflammatory, vascular, infectious, or neoplastic causes. Malignant small round blue cell tumors of the orbit are rare in adults and pose significant diagnostic challenges. Ewing's sarcoma, an aggressive malignant neoplasm of bone and soft tissue, is exceedingly uncommon in the orbit, particularly in adults. A distinct histological variant, adamantinoma-like Ewing's sarcoma (AL-EWS), demonstrates epithelial differentiation and adamantinoma-like features, confirmed by immunohistochemical positivity for CD99, cytokeratin, and FLI-1. [3,4]

Due to its rarity and overlapping features with benign orbital conditions, AL-EWS may be misdiagnosed, leading to delayed management. Early recognition is essential given its aggressive local invasion and potential for systemic spread. We present a rare case of AL-EWS in a 42-year-old female with progressive proptosis, emphasizing clinical, radiological, and histopathological findings, management, and short-term outcomes. [1,2]

MATERIALS AND METHODS

Study Design: Case Report

Study Setting: Department of Ophthalmology, RIO, MINTO OPHTHALMIC HOSPITAL, BANGALORE, a tertiary care centre.

Case Presentation

A 42-year-old female presented with complaints of swelling and outward protrusion of the left lower eyelid for the past 2 months. The onset was insidious, and the condition was gradually progressive. The patient reported associated diminution of vision and a pricking sensation in the left eye. She had a history of spectacle use for the past year. There was a history of restriction of ocular movements.

She denied any history of redness, watering, itching, discharge, diplopia, flashes, floaters, ocular trauma, or previous ocular surgeries. The patient had previously consulted at a private hospital, where she was prescribed oral steroids, which were discontinued abruptly after 3 days.

Clinical Examination

General Examination: Swelling of the left lower eyelid with outward protrusion of the left eyeball.

Table 1: Patient Demographics and Clinical History

Parameter	Findings
Age	42 years
Gender	Female
Duration of symptoms	2 months
Chief complaints	Swelling, outward protrusion of left lower eyelid
Associated symptoms	Diminution of vision, pricking sensation
Spectacle use	1 year
History of trauma	Absent
Previous ocular surgery	None
Prior treatment	Oral steroids (stopped after 3 days)

Table 2: Ocular Examination with Slit Lamp

	RIGHT EYE	LEFT EYE
VISUAL ACUITY	6/12-6/9	6/24-6/18
COLOR VISION	WNL	WNL
HEAD POSTURE	NORMAL	
OCULAR POSTURE	NORMAL,	AXIAL PROPTOSIS
LIDS	NORMAL	Swelling in the left lower eyelid
CONJUCTIVA	NORMAL	NORMAL
CORNEA	NORMAL	NORMAL
ANTERIOR CHAMBER	NORMAL DEPTH	NORMAL DEPTH
PUPILS	3mmRRR	3mmRRR
LENS	CLEAR	CLEAR
FUNDUS	WITHIN NORMAL LIMITS	WITHIN NORMAL LIMITS

Figure 1: Clinical Photograph Showing Axial Proptosis of the Left Eye.

Note the outward displacement of the left globe with prominence of the lower eyelid. No signs of periorbital inflammation or conjunctival congestion are evident.

Local Examination

- **Inspection:**
  - Left eye axial proptosis.
  - No visible pulsations or orbital swelling.
  - No lagophthalmos.
  - Skin over lids normal.
  - No evidence of periorbital inflammation.
  - No change in proptosis on bending forward or Valsalva manoeuvre.
- **Palpation:**
  - Findings confirmed.
  - No tenderness or local rise of temperature.
  - Finger insinuation test: Finger could be insinuated between globe and orbital rim.
  - Retropulsion test: Resistance felt on pushing globe backwards.
  - No thrill.
  - No supra-orbital or infra-orbital anaesthesia.
- Auscultation: No bruit.

Table 3: Local Examination Findings

Test/Observation	Findings
Inspection	Axial proptosis, no pulsations, no lagophthalmos
Skin over lids	Normal
Periorbital inflammation	Absent
Valsalva/Bending forward	No change in proptosis
Palpation	No tenderness, resistance on retropulsion
Finger insinuation test	Positive (space between globe and orbital rim)
Retropulsion test	Resistance felt
Thrill	Absent
Supra/Infra-orbital sensation	Normal
Auscultation	No bruit

Proptosis Evaluation

Quantitative assessment showed increased palpebral aperture and proptosis measurements in the left eye compared to the right: horizontal aperture 32 mm versus 28 mm, vertical aperture 12 mm versus 10 mm, axial proptosis 25 mm versus 20 mm, horizontal proptosis 27 mm versus 22 mm, and vertical proptosis 25 mm versus 20 mm. These findings confirmed significant unilateral proptosis of the left eye, supporting the presence of an orbital lesion.

Table 4: Proptosis Evaluation

Parameter	Right Eye	Left Eye
Palpebral Aperture (Horizontal)	28 mm	32 mm
Palpebral Aperture (Vertical)	10 mm	12 mm
Axial Proptosis	20 mm	25 mm
Horizontal Proptosis	22 mm	27 mm
Vertical Proptosis	20 mm	25 mm

Diagnosis

Magnetic resonance imaging (MRI) of the orbit revealed a well-defined soft tissue mass occupying the inferomedial extraconal space of the left orbit, causing anterior displacement of the globe and restriction of ocular movements. The lesion appeared heterogeneously enhancing, suggestive of a neoplastic process rather than an inflammatory or vascular pathology. In view of the progressive proptosis and radiological suspicion of malignancy, the patient was taken up for surgery. A left eye vertical lid-split anterior orbitotomy was performed, providing direct access to the anterior orbital compartment. Through this approach, careful debulking of the mass was achieved, minimizing damage to surrounding structures. Following surgical debulking of the orbital mass, the biopsy specimen was sent for histopathological and immunohistochemical evaluation. Microscopy revealed sheets of small round blue cells with adamantinoma-like features. Immunohistochemistry demonstrated positivity for cytokeratin (patchy granular), EMA, synaptophysin, chromogranin, NSE, CD99, NKX2.2 (diffusely strong), and P63 (patchy), while the tumor cells were negative for LCA, S100, desmin, and GFAP. Taken together, these immunomorphological findings were consistent with adamantinoma-like Ewing's sarcoma (AL-EWS), confirming the diagnosis in the biopsy of the retro-orbital mass of the left eye.

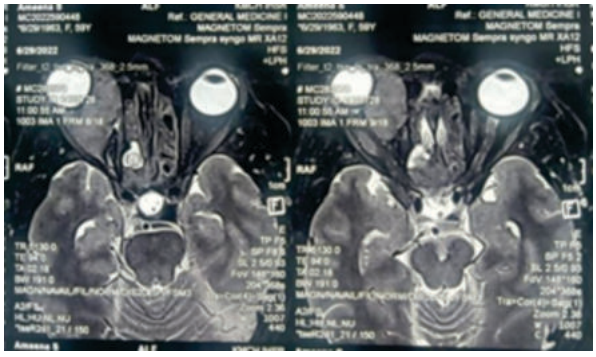

Figure 2: Axial MRI Scan of the Orbit Showing Retro-Orbital Mass

Axial T1-weighted MRI slices reveal a heterogeneously enhancing soft tissue lesion in the inferomedial extraconal compartment of the left orbit, causing anterior displacement of the globe. These findings support the presence of a neoplastic process.

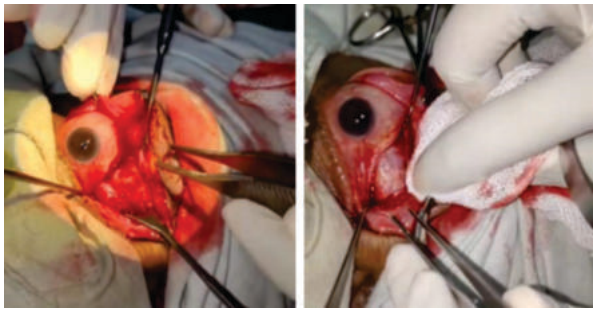

Figure 3: LE Vertical Lid Split Anterior Orbitotomy with Debulking of Mass was Done and Specimen Sent for Histopathology.

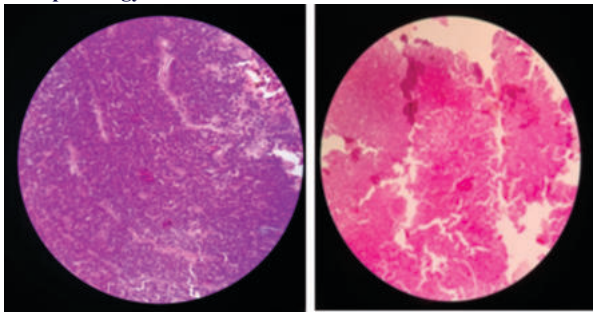

Figure 4: Histopathological Image of Retro-Orbital Mass (H&E Stain)

Haematoxylin and eosin-stained section showing sheets of small round blue cells with hyperchromatic nuclei and scant cytoplasm, consistent with adamantinoma-like features. These findings support the diagnosis of Adamantinoma-like Ewing's Sarcoma.

DISCUSSION

Adamantinoma-like Ewing's Sarcoma (AL-EWS) is a rare histologic variant of Ewing's sarcoma characterized by epithelial differentiation and small round blue cell morphology.[1] While classic Ewing's sarcoma typically affects children and adolescents, AL-EWS has been increasingly recognized in adults, often presenting in atypical locations such as the orbit. [1,2]

In this case, the patient presented with progressive unilateral proptosis, a symptom that overlaps with benign orbital conditions such as thyroid eye disease, idiopathic orbital inflammation, or cavernous haemangioma. [3] The absence of pain, redness, or systemic

symptoms initially masked the aggressive nature of the lesion. MRI revealed a well-defined, heterogeneously enhancing mass in the inferomedial extraconal space, raising suspicion for a neoplastic process. [4]

Histopathology showed sheets of small round blue cells with adamantinoma-like features. Immunohistochemistry was pivotal in confirming the diagnosis, with diffuse positivity for CD99 and NKX2.2, along with patchy expression of cytokeratin, EMA, and P63. [5] Negative markers such as LCA, S100, desmin, and GFAP helped exclude lymphoma, melanoma, rhabdomyosarcoma, and glial tumors respectively. [6]

AL-EWS is known for its aggressive behaviour and potential for systemic spread. Multimodal management—including surgical debulking, histopathological confirmation, and oncologic evaluation—is essential. Literature suggests that early diagnosis and complete excision, followed by chemotherapy or radiotherapy when indicated, can improve prognosis. [7]

CONCLUSION

Adamantinoma-like Ewing's Sarcoma should be considered in the differential diagnosis of progressive proptosis in adults, especially when imaging suggests a solid orbital mass. Its ability to mimic benign conditions underscores the importance of thorough clinical evaluation, advanced imaging, and immunohistochemical profiling. Prompt multidisciplinary intervention is vital due to its aggressive nature and potential for systemic dissemination.

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