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UNVEILING THE UNCOMMON: A CASE REPORT OF ROSAI DORFMAN DISEASE OF THE EYE.

Ophthalomology

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ABSTRACT

Purpose: To report a case of Rosai Dorfman Disease in right eye confirmed by MRI orbit and histopathological examination. **Method:** This case report describes a 65 year old female with a 7 year history of swelling and outward protrusion in her right eye who presented to Minto Ophthalmic hospital. **Result And Conclusion:** Complete detailed ophthalmic examination was done followed by MRI orbit and orbitotomy and debulking procedure. The specimen was sent for histopathological examination. Rosai Dorfman disease is a rare benign condition characterized by histiocytic proliferation and its manifestation in the eye is uncommon. It is essential to differentiate this condition from malignant histiocytosis as the treatment option differs.

KEYWORDS

Proptosis, Rosai Dorfman disease, Orbitotomy, MRI orbit, Histopathological examination

INTRODUCTION

- Rosai Dorfman disease (sinus histiocytosis with massive lymphadenopathy) is a rare, idiopathic, non neoplastic histioproliferative disease of unknown etiology.
- Most common extranodal sites include the eyes, ocular adnexa and orbit.
- Extranodal involvement occurs in 43% cases ,with ophthalmic disease observed in 11.5% of which orbital involvement is seen in 2.3% cases.
- Presently designated under R group histiocytosis, non Langerhans cell benign histiocytosis
- Most common presentation- painless lymphadenopathy with proptosis
- other symptoms- eyelid involvement, diplopia, headaches, lacrimal gland involvement, optic nerve compression in severe cases.
- The involved histiocytes express S-100 protein, HAM 56, α1 antitrypsin, α1 chymotrypsin, lysozyme, Mac 387, Ki-1 but are negative for Cd1a

Case Description

- A 65 year old female presented to oculoplasty clinic with complains of swelling and outward protrusion of right eye since 7 years. It was insidious in onset and gradually progressive.
- Also c/o pain, pricking sensation and watering.
- H/o cataract surgery in RE 10 years back. Patient had previously undergone debulking procedure twice in the past 7 years and was diagnosed as Rosai Dorfman disease as per histopathological report.
- General physical examination showed enlarges submandibular lymph nodes on the right side.

Ocular Examination

- **Head Posture-** normal
- **Ocular Posture-** Abaxial proptosis, right eye pushed down and outward

Table 1: Ocular Examination Of The Patient

	RIGHT EYE	LEFT EYE
Vision	6/60 6/24	6/24 6/18
Lids	Edematous, superonasal fullness	normal
Conjunctiva	congested	normal
Cornea	clear	Clear
Pupil	Grade 1 RAPD	3mm, RRR
Lens	PCIOL	SIMC
EOM	-3 -4 / -2 -4 / -2 -4	N N / N N / N N
Fundus	Grade IV disc edema with tortuous vessels	Wnl

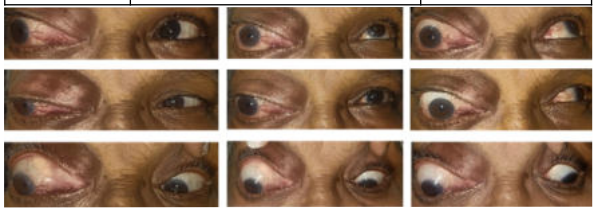

Image 1: 9 gaze photos

Proptosis Evaluation

- Abaxial proptosis with superonasal fullness seen in RE.
- Swelling measuring 7X5 mm noted in superonasal quadrant with diffuse borders, firm in consistency, non mobile, tender.
- Resistance felt on retropulsion test

Table 2: Proptosis Evaluation Of The Patient.

	RE	LE
Horizontal palpebral aperture	38mm	28mm
Vertical palpebral aperture	14mm	10mm
Axial	26mm	15mm
Horizontal	33mm	25mm
Vertical	32mm	25mm

Image 2: Image showing abaxial proptosis

Mri Brain And Orbit

- s/o well defined multi lobulated heterogeneously enhancing soft tissue intensity mass lesion in intraconal space of right orbit displacing optic nerve laterally and abutting posteromedial aspect of globe, medial rectus and superior oblique muscles. Heterogenously enhancing soft tissue thickening in right maxillary sinus with osseous changes in walls of paranasal sinuses: all s/o histiocytic disorders like Erdheim- Chester disease or Rosai Dorfman disease

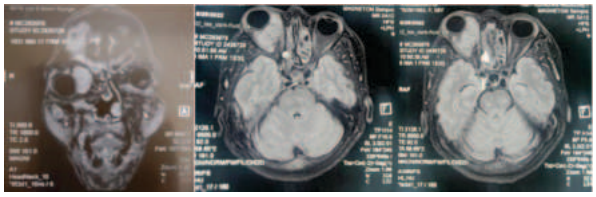

Image 3: MRI orbit images of the patient showing right eye proptosis

Management

- RE vertical lid split anterior orbitotomy with debulking of mass was done and specimen sent for histopathology

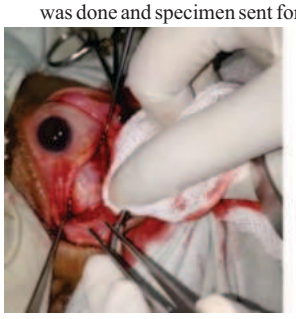

Image 4: Intraoperative image

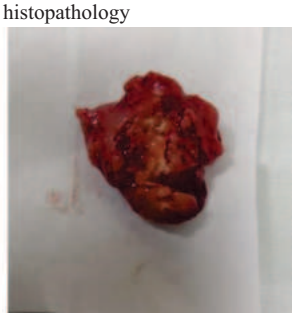

Image 5: Specimen sent for HPE

- Histopathological impression- sheets of foamy histiocytes with admixed dense plasma cells and mononuclear infiltrates. Numerous Russel bodies present. PAS- negative, Perl's stain- Negative

Post Operative

Image 6: Post operative image

DISCUSSION

- Clinical diagnosis is difficult and exact pathogenesis is unclear. Some studies have shown that RDD may be related to human herpesvirus 6 and Epstein-Barr virus infection.
- Imaging like MRI is essential for evaluation
- Definitive diagnosis is by histopathological examination and molecular studies
- May recur and require multiple and repeated debulking surgeries

CONCLUSION

- Rosai-Dorfman disease may be suspected in individuals with unilateral or bilateral slowly progressive proptosis with or without massive cervical lymphadenopathy.
- Systemic evaluation is needed to document other extranodal sites of involvement

This case illustrates the importance of histopathological analysis in accurate diagnosis as it is necessary to differentiate it from neoplastic etiologies like Hodgkin lymphoma, Langerhans cell sarcoma

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