Mature Orbital Teratoma
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ABSTRACT
Orbital teratoma is a rare tumour that is composed of tissues derived from all three germ cell layers. It presents, in otherwise, healthy neonates with unilateral proptosis that progresses rapidly, threatening vision through optic nerve compromise or exposure keratopathy. Globe conservation may be difficult too in advanced cases. Mature teratomas are benign, but the immature ones can sometimes become malignant. We report a case of a six-month female infant who presented with mature orbital teratoma since birth. Vision could not be salvaged; but following complete surgical excision, good cosmesis was achieved.

Key Words: Mature teratoma, Orbit, Congenital tumour, Surgery.

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INTRODUCTION
Congenital orbital teratomas are rare tumours composed of tissues derived from all three germinal layers.\(^1\) They cause severe unilateral proptosis that may result in optic nerve compromise and exposure keratopathy.\(^2\) They show rapid growth but malignant conversion is rare.\(^3\) Most common locations for teratoma are gonads, sacrococcyx, retroperitoneum and mediastinum.\(^3,4\) Head and neck teratomas make up only 9% of all cases and among these, the orbital teratomas make up a very small number.\(^3,4\) Recurrences can occur.\(^5\) The aim of management is primarily to save the globe so that normal orbitofacial development is possible. Salvaging the vision is not possible in many cases due to secondary optic atrophy and exposure keratopathy. Here, we report a case of a six-month female infant, who presented with a mature orbital teratoma that had compromised the vision due to secondary optic atrophy, but the globe could be saved and good cosmetic outcome achieved.

CASE REPORT
A six-month female infant, result of a non-consanguineous marriage and uneventful term delivery, presented with progressive proptosis of left eye since birth. On examination, extreme proptosis and lateral displacement of the left globe along with distension of lids was seen (Figure 1). Mild conjunctival hyperemia was seen but the cornea did not show any signs of exposure keratopathy. There was scarring along medial aspect of orbit, indicative of a surgical procedure in the past.

On further enquiry, it was revealed that a previous incisional biopsy had been done elsewhere; but the parents had lost the histopathology report. The child could only follow light in the left eye. Left relative afferent pupillary defect was present. Fundus examination showed pale optic disc in the left eye. The right eye was completely normal. The child was, otherwise, fully healthy.

Computed tomography scan (Figure 2) and magnetic resonance imaging revealed a huge well-defined largely cystic, multiseptated mass, filling up the left orbit completely, causing forward and lateral displacement of the globe. Expansion of bony orbit without any bony erosion was present. No extension to surrounding structures was seen. Optic nerve and extraocular muscles could not be visualised separately from the mass. Routine blood tests, chest x-ray and ultrasound abdomen were normal. Serum alpha fetoprotein (AFP) was elevated at 144.8
Complete surgical excision through a superomedial orbitotomy approach was performed. Globe was not penetrated by the mass so it was preserved. Histopathological examination revealed a lesion composed of mature, columnar epithelium, fibroadipose tissue, glial tissue, mucous and serous acini and mature cartilage. On the basis of the presence of mature elements, derived from all three germ cell layers, diagnosis of mature orbital teratoma was made. Although vision could not be salvaged in our patient because of late presentation, but good cosmetic outcome was achieved (Figure 3). Six-monthly follow-up was advised to look out for recurrence.

CONCLUSION

Orbital teratomas are very rare, yet potentially curable congenital tumours, so they should be included in the differential diagnoses of rapidly progressive proptosis in a neonate. Moreover, early surgical intervention aiming at compete surgical excision with globe preservation, wherever possible, should always be considered.

PATIENT’S CONSENT:
Formal consent was taken from the parents of the child to publish the data related to the case.

CONFICT OF INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
RC, MNQ: Diagnosis and management.
AR: Data collection and manuscript writing.

REFERENCES


