

## Case report

### Orbital Teratoma: A rare congenital tumour

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#### Abstract

**Background:** Orbital teratomas are congenital, unilateral germ cell tumors, which are present at birth with moderate to massive proptosis. The rare tumour has to be managed individually and is at times difficult. **Case:** A female child in her 2<sup>nd</sup> day of life was brought to our department with complains of forward bulging of the left eye which was noticed since the time of her birth. **Observation:** Examination and investigations revealed a mass with scattered foci of calcification. The mass was causing expansion of left orbit with thinning and scalloping of bony boundaries of left orbit with no intracranial component. The scan suggests the diagnosis of orbital teratoma which was further confirmed by histopathological examination. **Conclusion:** The rare tumour presents a challenge in management. The aim of management depends upon the extent of tumour, preservation of vision whenever possible, promotion of normal orbital growth and cosmesis.

**Keywords:** Orbital teratoma, rare, neonate

#### Introduction

Congenital tumours of the orbit or elsewhere may be derived from one, two, or all three germ cell layers and are named accordingly (Bernoulli, 1962). The word "teratoma" means "monstrous growth" in Greek. These tumors are the rarest of congenital orbital tumors and constitute all three germ cell layers which are arranged in varying degrees of complexity (Simonsen et al, 1981). However, teratomas usually take the form of a shapeless mass without a regular tissue configuration (Ferry, 1965).

#### Case Report

A female child in her 2<sup>nd</sup> day of life was brought to our emergency department by her parents from Pyuthan with complain of forward bulging

of left eye which they noticed since the time of her birth. The swelling was progressive. The estimated size of the swelling was about that of a tomato at birth which progressed rapidly to become the size of a small coconut before they sought any medical attention. They initially went to nearby district hospital and then to Bhairahawa and finally referred to our centre.



**Figure 1:** The child with left orbital mass at the time of presentation.

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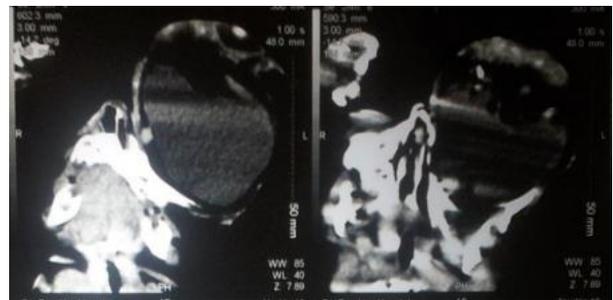
Upon inquiry she was born to a healthy 21 years old mother following an uneventful pregnancy and delivery events and with no other systemic abnormalities. On clinical examination, the child was alert and stable with normal breathing and colour. Ocular examination showed upwardly displaced left eyebrow, stretched eyelid and keratinized conjunctiva. However, other details of the globe regarding anterior and posterior segment as well as the fundal glow couldn't be appreciated. Upon examination of the mass, it was extending beyond the orbital rim, reddish, globular, smooth, with no rise in temperature, measuring 5\*5\*5 cm, variable (cystic to firm) in consistency, absent pulsation, positive fluctuation, partial illumination and no bruit on auscultation. With co-ordination from pediatrics and neurosurgery departments, the child was admitted in our ward for further management.



**Figure 2:** The orbital mass is seen pushing the globe anteriorly (note the keratinized cornea)

Her CT scan of head and orbit showed a large heterogeneous mass measuring 7\*6.5\*5 cm in the retro-bulbar region of the left globe which was displacing the globe anteriorly. The lesion had fluid attenuating component +23 HU posteriorly and fat attenuating component anteriorly -60 HU with fat fluid level. The mass had scattered foci of calcification and causing expansion of left orbit with thinning and scalloping of bony boundaries of left

orbit. However there was no extension into the intracranial component. Left optic nerve and extra-ocular muscles were not separately visualized. The study of brain was normal. Also USG Doppler of left orbit revealed large space occupying lesion in left orbit measuring 4\*4 cm with vascularity and displacing the globe antero-superiorly with foci of calcification and minimal cystic component within the lesion. Finally MRI of brain and orbit was done as per the advice of radiologist which divulged the similar findings as that of CT scan. All these imaging indicated a possibility of orbital teratoma.



**Figure 3:** CT scan showing heterogenous mass which is compressing the globe and expansion of left orbit.

The child was then scheduled for elective enucleation with complete removal of the tumour. The details of a challenging surgery performed on a fragile neonate: the orbital mass was examined in detail for its extent. Horizontal conjunctival incision was given on either side of keratinized conjunctiva and extended. The mass was completely excised (in toto) after dissecting from other orbital tissues. Optic nerve was identified and resected after cauterization. Orbital rim palpated and prominent lateral orbital rim (? cartilaginous overgrowth) was excised and smoothed. The conjunctiva was sutured and only appropriate sized conformer was placed, however ball implant was not implanted owing to the fact of large orbital cavity. Lastly upper and lower lid tarsorrhaphy executed and eye padded.



**Figure 4:** Excision of the mass in toto.

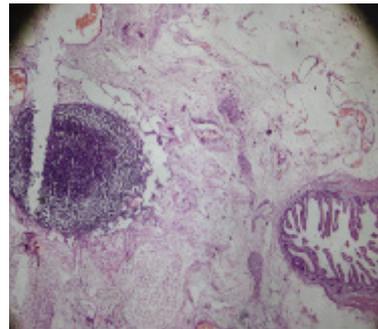
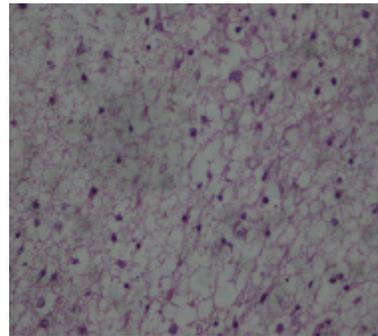
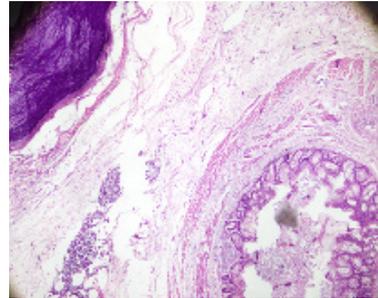


**Figure 5:** Compressed globe after excision of the mass (note the intact optic nerve)

Two different specimen samples were sent: one that of the mass and the other of the compressed eyeball. The dispatched histopathological report revealed cystic structure lined by cuboidal epithelium. There were also cartilages, lymphoid follicles, mature glial cells, mature adipose tissues, smooth muscles and some glands lined by colonic type of mucosa. The final histopathological findings vouched for the diagnosis of mature teratoma.



**Figure 6:** Postoperative picture of the child before discharge



**Figure 7:** Histopathological slides showing intestinal mucosa, neural elements and lymphoid follicles.

### Discussion

Teratoma is a congenital tumour of cystic nature and represents all of the three embryonic germ layers: ectoderm, mesoderm, and endoderm histologically. Due to its rarity, orbital teratomas are usually cited as case reports individually in the literature (Shields et al, 2004). Teratomas, the most common of orbital germ cell tumors, probably arise from pluripotential embryonic stem cells that are carried to the orbit by blood circulation and escape regulatory influences, or from primordial germ cells that aim towards the pineal gland, even though other theories have also been proposed (Gonzalez, 1982).

A primary orbital teratoma presents at birth as a large mass causing moderate to massive proptosis, distended eyelids, chemotic conjunctiva and sometimes keratinized due to drying. They preferentially affect females and with equal frequency of laterality. Most tumors are intraconal and stretch the four recti, which results in a quadrangular shape and axial proptosis. The eye might be buried within the tumor itself, and only the cornea or a narrow rim of the sclera visible. Exposure keratopathy, ulceration, keratoiritis, and even spontaneous perforation occur secondarily (Tero et al, 1994).

Imaging characteristically shows a well-delineated heterogeneous mass with cysts, punctate calcification, and areas of fat density (Kaufman et al, 1998). The clinical management of these lesions is unclear, due in part to their low incidence and to an incomplete understanding of their natural history. Surgical excision may be curative without affecting function (Lee et al, 1997).

The prognosis of teratoma is related to several factors including the age of the patient, the site, and immaturity of the tumor. In the past, complete excision of orbital teratomas has been advocated early in neonatal life. But when the eye is normal but damaged by pressure from the tumor, lid-sparing exenteration may be recommended (Bilgic et al, 1997). It is hard to recommend a fixed management plan, however a common agreement in the management objectives should be to save the eye, retain some vision, encourage normal orbitofacial development, and maintain cosmesis (Gnanaraj et al, 2005).

### Conclusion

Orbital teratoma is a rare, benign but disfiguring congenital tumour of the orbit. It is imperative to rule out its intracranial extension and complete excision of the tumour is mandatory. The preservation of vision should be aimed for

whenever possible and also measures taken for normal orbital development.

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