

## Congenital orbital teratoma

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We present a case of mature congenital orbital teratoma managed with lid-sparing exenteration and dermis fat graft. This is a case report on the management of congenital orbital teratoma. A full-term baby was born in Fiji with prolapsed right globe which was surrounded by a nonpulsatile, cystic mass. Clinical and imaging features were consistent with congenital orbital teratoma. Due to limited surgical expertise, the patient was transferred to Adelaide, Australia for further management. The patient underwent a lid-sparing exenteration with frozen section control of the apical margin. A dermis fat graft from the groin was placed beneath the lid skin to provide volume. Histopathology

revealed mature tissues from each of the three germ cell layers which confirmed the diagnosis of mature teratoma. We describe the successful use of dermis fat graft in socket reconstruction following lid-sparing exenteration for congenital orbital teratoma.

**Key words:** Congenital, dermis fat graft, orbital, teratoma

We present a case of mature congenital orbital teratoma managed with lid-sparing exenteration and dermis fat graft.

### Case Report

A full-term baby girl of 3.5 kg was born with massive right proptosis [Fig. 1]. Antenatal progress was normal and no intrauterine ultrasound was performed. She was otherwise well and the delivery was unremarkable. On examination, there was complete right globe prolapse associated with restrictive ophthalmoparesis. The globe was surrounded by a nonpulsatile, cystic mass which transilluminated [Fig. 1]. Exposure keratopathy and severe anterior chamber inflammation with 3 mm of hypopyon was present. There was an absolute afferent pupillary defect in the affected eye. There was no view of the fundus. The left eye and the rest of clinical examination was normal. The eye was lubricated and covered with cling wrap while awaiting intervention.

B-scan ultrasound demonstrated a heterogeneous, multicystic mass. Computerized tomography (CT) demonstrated calcification of the lesion and globe displacement anteriorly and the optic nerve laterally [Fig. 2]. The orbit was expanded but there was no intracranial invasion. An incisional biopsy revealed fibroadipose tissue with areas of differentiated colonic mucosa with goblet cells and submucosal lymphocyte aggregates (Peyer's patch). The histology was consistent with mature orbital teratoma.

At 7 weeks, the baby was transferred to Adelaide, Australia for further management. While awaiting transfers, the cystic component grew in size and required aspiration of the anterior

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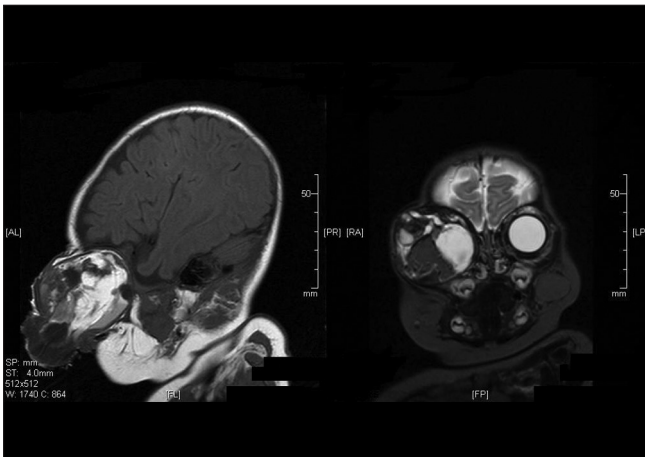
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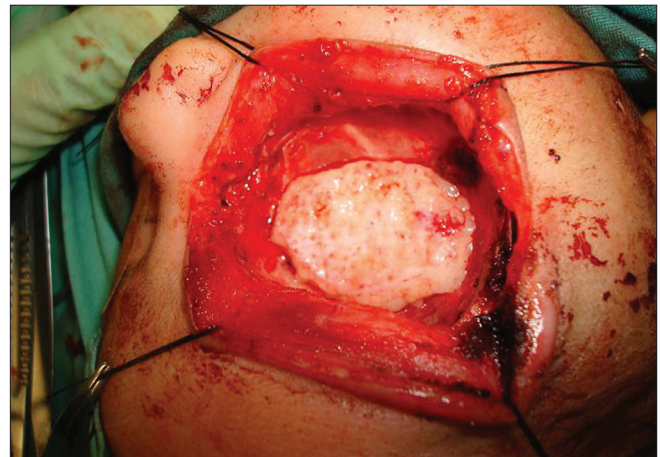
**Figure 1:** Preoperative image demonstrating large, cystic mass with nonviable eye



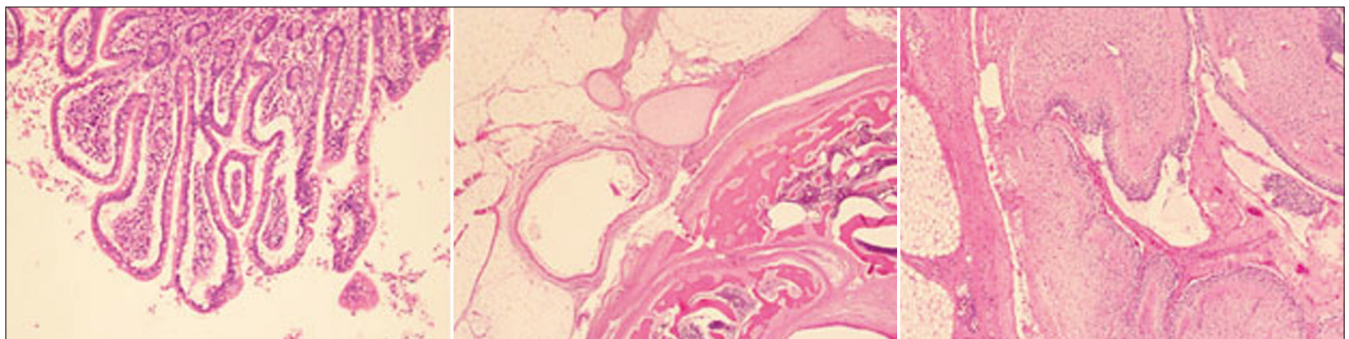
**Figure 2:** Computerized tomography (CT) of the orbit showing punctate calcification of the lesion and anterior globe displacement



**Figure 3:** Magnetic resonance imaging of the head and orbits showing an irregular, heterogeneous mass encasing the optic nerve and the globe



**Figure 4:** Exenterated orbit with dermis fat graft *in situ*



**Figure 5:** Micrograph of representative mature tissues derived from the three embryonic germ cell layers: Small intestine derived from endoderm (far left) bone and cartilage derived from mesoderm (centre) and cerebellum derived from the ectoderm (far right)

cysts. Magnetic resonance imaging and CT angiography confirmed no intracranial or sinus extension and no major vascular connection apart from a normal caliber ophthalmic artery [Fig. 3]. She underwent a lid-sparing exenteration with

frozen section control of the apical margin. A dermis fat graft from the groin was placed beneath the lid skin to provide volume [Fig. 4]. There was no intraoperative complication and the postoperative course was uncomplicated. Histopathology



**Figure 6:** Appearance at 2 weeks postoperation



**Figure 7:** Appearance at 8 months of age

revealed mature tissues from each of the three germ cell layers. The teratomatous elements include salivary gland, thyroid, cerebellum, gastrointestinal mucosa, skin, bone, cartilage, adipose tissue, and skin adnexal structures [Fig. 5]. Some of the epithelial tissues had formed variably dilated cystic spaces. No immature elements were identified thus confirming the diagnosis mature teratoma.

The patient returned to Fiji 2 weeks following surgery [Fig. 6] and was doing well at 6 months postoperation [Fig. 7]. Yearly follow-up was planned to assess for the growth of socket and to ensure there are no recurrence.

## Discussion

To the authors' knowledge, this is the first reported case in Fiji. The surgical expertise and investigations required lead to transfer of the baby to from Fiji to Australia.

Orbital teratoma is a rare but important differential for proptosis in a newborn.<sup>[1,2]</sup> By definition, to make the diagnosis of congenital orbital teratoma, tissues from all three embryonic germ cell layers, i.e. ectoderm, mesoderm and endoderm must be present.<sup>[1,2]</sup> Teratoma are considered mature if the tissue present in the teratoma are fully differentiated and does not contain evidence of immature embryonal tissue.<sup>[2]</sup> Teratoma is distinct from dermoids which consists of a single germ cell layer and teratoid which consists of 2 germ cell layers.<sup>[1]</sup> Complication of teratoma includes cranio-orbital invasion that can be fatal and if incompletely excised, recurrence or even malignant transformation may occur.<sup>[2]</sup>

It is well described that removal of the eye or orbital contents in an infant will lead to impaired bony development of the orbit and ipsilateral facial bones.<sup>[3]</sup> Hence, a dermis fat graft was used to provide volume and the stimulus to bony growth.<sup>[3]</sup> As the orbital volume was 50% larger compared with

the contralateral side, it is anticipated that there will not be a significant asymmetry in the long term. Alternatives would have included a ball implant, a temporalis flap or a free flap. As preoperative embolization has been utilized in orbital teratomas to decrease intraoperative blood loss,<sup>[4]</sup> the patient underwent a CT angiogram but this did not show significant vascularity it was felt that exenteration could proceed without embolization.

Although preservation of the globe has been successfully performed for selected cases of teratoma,<sup>[1,2]</sup> the extent of tumor, severe compression of the globe, and the inability to reliably isolate the recti and globe from the tumor lead to an eyelid-sparing exenteration. Intraoperatively, it was also apparent that the majority of the conjunctiva could not be dissected free of the tumor excluding the possibility of a conjunctiva sparing exenteration. In summary, we present a case of mature congenital orbital teratoma managed with lid-sparing exenteration and a dermis fat graft.

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