CASE REPORT

Congenital Orbital Teratoma with Unilateral Proptosis
Rubina Gulzar, Ruqaiya Shahid and Talat Mirza

ABSTRACT
Teratoma is a rare tumor, containing structures originating from all three germinal layers. The most frequent location of teratoma is the gonads. Orbital teratoma is extremely rare. In 1969, Jensen reported that only 40 cases of orbital teratoma existed in the world. We describe a rare case of orbital teratoma in a 15-day girl who presented with marked proptosis of the left eye. The eyeball was embedded within the mass that could be seen all around the globe. Orbitectomy was performed with the impression of retinoblastoma. Histopathological examination was reported as benign mature cystic teratoma; no immature component was identified. The case is being reported because of its rarity and also to highlight the use of modern neuroimaging techniques in making an accurate preoperative diagnosis, which helps in better operative management of these patients.


INTRODUCTION
The word "teratoma" means "monstrous growth" in Greek.1 Teratoma comprises 6.6% of all childhood tumors.2 It is composed of derivatives of all three germinal layers, arranged in a haphazard manner. It originates from totipotential germ cell, which has the capability to differentiate into all three germinal layers.3 Congenital teratoma is usually gonadal. It can also be observed in the sacrococcygeal and retroperitoneal regions and tends to have a favourable outcome, if promptly excised.3

CASE REPORT
The patient was seen at the age of 15 days for proptosis of the left eye, since birth. Details of the pregnancy, birth, and family history were unremarkable. On ophthalmologic examination, there was severe proptosis of left eye: the globe was extending beyond the orbital confines with corneal ulceration. The swelling was variably hard and soft in consistency and non-tender. Fundoscopic examination revealed a normal sized vasculature, sharp discs in both eyes, and scattered blot hemorrhages in the affected eye. Routine hematological and biochemical laboratory tests were within normal limits. As the patient belonged to a low socioeconomic status and had visited a government hospital with scarce resources, magnetic resonance imaging (MRI) studies were not performed.

The patient underwent left modified eye exenteration, on the clinical impression of retinoblastoma, on the 20th day of her life. The distorted globe and a well encapsulated orbital tumor were completely removed. No postoperative complications were noted.

On gross examination, the intact, complete specimen measured 5 x3.5x 2cm. The cut section showed that the eyeball, though encased by the tumor, was delineated from it. The mass was well-encapsulated. It had a greyish solid surface with multiple variable sized cystic spaces.

Microscopic examination revealed centrally located eye, comprising of well differentiated retinal epithelium, surrounded by the lesion derived from all three germ layers. The solid areas of tumor comprised of mature neuroglial tissue. Surrounding areas showed presence of fat, smooth muscle, nerve bundles, lymphoid tissue, islands of cartilage and bone (Figures 1 and 2). The cysts were lined by squamous, columnar and respiratory mucosa. No immature component was identified. A diagnosis of mature cystic teratoma was made.

Department of Pathology, Dow International Medical College, Dow Diagnostic Research and Reference Laboratory, Dow University of Health Sciences, Karachi.
Correspondence: Dr. Ruqaiya Shahid, Assistant Professor, Department of Pathology, Dow International Medical College, Dow Diagnostic Research and Reference Laboratory, Dow University of Health Sciences, Karachi.
E-mail: ruqaiyashahid@yahoo.com

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Figure 1: Low power view of teratoma, showing a cyst, smooth muscle, adipose tissue and cartilage.
DISCUSSION

The first case of orbital teratoma was reported by Holmes in 1862.4 Orbital teratoma can primarily arise in the orbit or intracranial teratoma and invade the orbit secondarily. Congenital orbital and intracranial teratomas are uncommon.5 Bibliographic reviews refer a slight predominance of the left orbit (60%) and a male to female ratio of 1:2.6 Clinical presentation is proptosis of the affected eye, in an otherwise healthy newborn, with distended eyelids, chemotic conjunctiva, exposure keratopathy, and rapid growth of the tumor after birth.5 Tumors untreated for months can achieve larger size and, therefore, early diagnosis is critical. Complications include corneal ulceration, optic atrophy, and facial deformities.7 Rare case of malignant germ cell tumor, arising in benign orbital teratoma, has been reported.7 Teratoma must be distinguished from conditions such as congenital glaucoma, hematoma, retrobulbar hemorrhage, benign rapidly growing orbital tumors such as, hemangioma, lymphangioma, meningoecele, encephalocele and malignant tumors such as rhabdomyosarcoma, neuroblastoma and leukemia.8,9 A diagnosis of orbital teratoma should be considered due to the presence of unilateral proptosis, expansion of the orbit, fluctuation, and trans-illumination of the mass.1 Radiologic studies are essential for a prompt diagnosis. MRI is the standard criterion and is usually performed with a CT scan. The most common radiologic finding is the presence of a heterogeneous cystic mass with varying signal intensity and often with calcification.2 There may be an enlargement of the orbital bone with deformation of facial bones.2 Unfortunately, in this case radiological studies were not performed. The treatment of choice is complete tumor excision with sparing of the eye, to maintain some vision, whenever possible.10 In the past, many surgeons preferred orbital exenteration because of possibility of malignancy.4

In conclusion, congenital orbital teratoma is potentially curable and must be considered in the differential diagnoses of early childhood orbital tumor.

REFERENCES


Figure 2: Teratoma showing lobules of cartilage, bone formation, fat, smooth muscle and a cyst with respiratory epithelium.