

Congenital Orbital Teratoma: A Case Report

Abstract

Teratomas are germ cell tumors that may contain several tissues derived from one or more of the three germ layers. Congenital orbital teratomas are rare. We report a case of congenital orbital teratoma in a 7-h-old male neonate who was noticed to have periocular swelling and progressive proptosis of the left eye at birth. Orbital teratoma was suspected based on the clinical presentation and imaging findings. Treatment was conducted by modified exenteration, and the diagnosis was confirmed histologically as mature teratoma.

Keywords: *Congenital proptosis, exenteration, orbital teratoma, prosthesis, zaria*

Key Messages: *Congenital orbital teratomas may be rare but still remain a significant differential diagnosis of neonatal proptosis.s*

Introduction

Teratoma means “monstrous growth” in Greek. Originating from totipotential cells, it is composed of tissues derived from all three fetal germinal layers, which are arranged in a haphazard manner.^[1,2] A teratoma is different from a dermoid, which has a single germ cell layer, and a teratoid, which consists of two germ layers. Teratomas are typically found in the midline; most commonly in the sacrococcygeal region (40%); and finally, in the gonads, cervical, and retroperitoneal regions of the body.^[3] Teratomas are uncommon in the head and neck regions.^[4] Orbital location is rare (0.8%)^[3] and it usually occurs in the left orbit of healthy newborns, with a female: male ratio of 2: 1.^[5] In Nigeria, only four cases have been reported till date. Clinically, the tumors present with rapid growth, unilateral proptosis, and palpebral retraction without intracranial involvement.^[6]

Case History

A 7-h-old boy was referred to the eye clinic by the neonatologist, with forward protrusion of the left eye noticed at birth. There was associated periocular swelling, redness, incomplete closure of the eye, and no bleeding. The boy was delivered by spontaneous vertex delivery at full term (41weeks and 3days). The mother was healthy throughout pregnancy but was told she had excessive amniotic fluid

on an ultrasound scan during one of her antenatal care visits. The boy was the sixth child of the family, and all the older siblings were well. There was no history of congenital abnormalities in the family or among the relations.

Examination revealed an active and healthy-looking male neonate who was not in any obvious distress. He weighed 4.5kg and had an occipitofrontal circumference of 34 cm. Both the anterior and posterior fontanelles were patent, pulsating, and of normal tension. There was a 2.5 mm axial proptosis of the left eye that did not increase with crying, did not pulsate, and was not reducible. The surrounding eyelid skin was of normal color, had no differential warmth and no mass was palpable. The eyelids were widely parted with a palpebral height of 9 mm [Figure 1]. There was inferior conjunctival chemosis, inferior cornea haze, and normal anterior chamber depth. No other abnormalities were found in the left eye. The right eye was essentially normal. No abnormalities were found in the other systems. Conservative treatment with artificial tears and protective dressing was commenced while awaiting the results of investigations.

Ocular ultrasound scan showed an ill-defined heterogenous hyper-echoic soft tissue, a retrobulbar mass with cystic components in the left orbit. The left globe was displaced anteriorly with no visualized optic nerve. Color Doppler interrogation showed no blood flow. Transfontanelle ultrasound

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scan showed normal brain tissue and a ventricular system. Magnetic resonance imaging (MRI) showed a large mass occupying the left orbit, displacing the globe anteriorly, with fluid contained within the mass and a remodeled orbital wall; otherwise, normal cerebral hemispheres, sulci and ventricles, and a normal brainstem were seen [Figure 2].

Blood tests, including full blood count, electrolytes, urea, and creatinine, were all within normal limits. Serum alpha fetoprotein and fine needle aspiration biopsy for cytology were planned but not done due to a lack of facilities. The proptosis progressed rapidly to 7 mm with keratopathy, thinning of the cornea, and uveal prolapse at 7–8 o'clock position despite the

application of antibiotic ointment and constant padding of the eye [Figure 3].

A modified exenteration of the left eye and orbit, sparing the eyelids, was performed at six weeks of life. A solid to cystic retrobulbar mass measuring 4 × 6 cm was excised with the eyeball and sent for histology. The child was placed on oral analgesics and antibiotics postoperatively.

On histopathological examination, macroscopically the tumor consisted of multiple irregular-shaped gray white tissue that aggregated to 5 × 4 × 2 cm and weighed 22 gm. Cut sections of the globe showed empty anterior and posterior compartments, whereas sections of the fragments showed gray brown solid areas. Microscopically, the tumor was composed of an admixture of different tissue types, including mature neural tissue, intestinal and respiratory type of glands, sebaceous and sweat glands, and skeletal muscles. Immature or malignant tissue was not identified. The conclusion was mature cystic teratoma (Grade 0, benign; based on the grading system for extragonadal teratomas).^[7,8]

The patient was discharged after two weeks with a clean socket [Figure 4] and referred to the oculoplastic unit, where orbital reconstruction and fixing of prosthesis was subsequently done for the patient.

Discussion

Orbital teratoma is a rare tumor that most often presents at birth. The first case was reported by Holmes^[9] in 1862, and



Figure 1: Proptosis of the left eye with chemosis

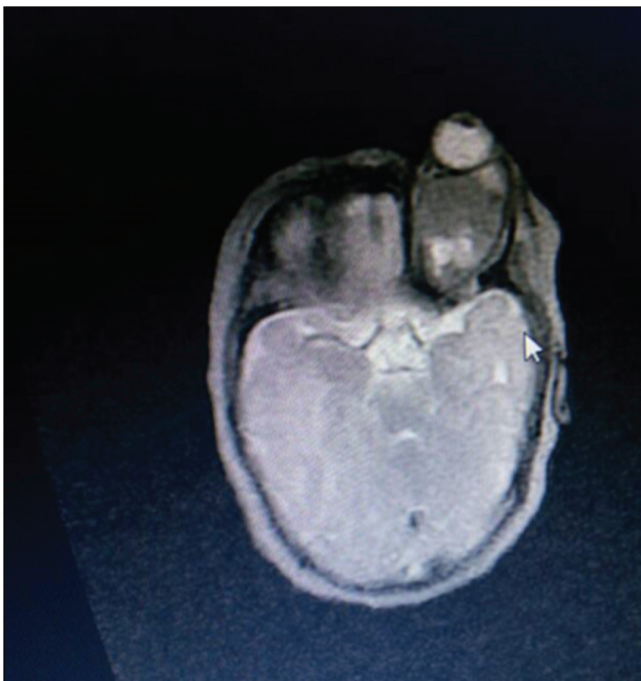


Figure 2: Brain MRI of the patient



Figure 3: Clinical photograph showing left proptosis and exposure keratopathy



Figure 4: Clean left orbital socket at discharge

approximately 70 cases have been reported worldwide.^[10] In Nigeria, very few cases have been reported till date. In 1999, Ameh *et al*^[11] reported the first case in Zaria. In 2000, Akabe *et al*^[12] reported a case with malignant transformation in Jos. A third case was reported from Onitsha in 2010 by Onyekwe *et al*.^[13] An immature orbital teratoma (Grade 1, probably benign) reported in Ibadan by Ogun *et al*^[8] in 2012 was similar to that reported by Ameh *et al*.^[11]

To the best of the authors' knowledge, this is the first case of mature cystic orbital teratoma to be reported in Nigeria. This is quite different from the previously reported cases due to the absence of immature or malignant tissue and its complete benign nature (Grade 0). Grades 1, 2, and 3 teratomas have <10%, 10–50%, and >50% amount of immature neuroectodermal tissue, respectively.^[7]

A few reports have suggested that very rarely, a benign teratoma may recur as a malignant germ-cell tumor. Orbital teratomas have been known to recur and may undergo malignant degeneration. Therefore, close follow-up is necessary.^[14]

The majority of the congenital orbital teratomas exhibit benign biologic behavior and are now being managed more conservatively with good cosmetic results and a better prognosis for vision.^[8] In some instances, as in the reported case, exenteration is necessitated by severe proptosis and irreversible damage to the globe. Modified and radical exenteration surgery could be disfiguring; the patients need early oculoplastic intervention to achieve favorable cosmetic outcomes. The lack of stimulation in the anophthalmic socket creates an imbalance for hemifacial and orbital development. This poses a challenge, as review surgeries are often required.^[15]

In conclusion, congenital orbital teratomas may be rare but still remain a significant differential diagnosis of neonatal proptosis. Early diagnosis and prompt management are necessary for a good outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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