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Case Report

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Beyond Belief: A Rare Encounter with Eye Teratoma

Vineet Shah*, Santosh Singh Patel and Rajesh Sahu

PT JNM Medical College, Raipur, Jail Rd, Opp Central jail, Moudhapara, Raipur, India

***Corresponding Author**
Vineet Shah, 102 Ankur Dadabhai Road, Andheri West, India, Tel: 9029690126.

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Abstract

A 7-year-old boy presented with progressive swelling in left eye since childhood. Examination revealed a soft cystic mass in the medial aspect of the left orbit. Rudimentary eye structures were present in left eye. There was no evidence of any exposure keratitis. A clinical diagnosis of congenital cystic eyeball was made and patient underwent cystic fluid aspiration followed by intralesional steroid injection followed by enucleation. Histopathology showed mature neuroglial tissue, fibro collagenous and vascular proliferation with mature cartilage suggestive of congenital mature teratoma. This case report tends to highlight the features of this rare case of congenital mature teratoma of eye.

Keywords: Cystic, Enucleation, Retinoblastoma, Teratoma

Introduction

Teratomas are tumors that are composed of different tissues derived from all three germ layers [1]. Orbital teratomas are rarely seen (0.8%) and usually present in the left orbit, with a more female preponderance [2,3]. Clinically, the tumors present with rapid progressive growth, proptosis usually unilateral and without intracranial involvement [4]. These may cause orbital enlargement to about three times its normal size [5]. On imaging, benign orbital teratomas are usually multiloculated cystic masses with a mixture of adjoining tissues which may include calcification, fat and ossification. Most orbital teratomas are sharply circumscribed, benign and well differentiated on histology [6].

Case Report

A 7-year-old boy presented with a progressive swelling in left eye since childhood. Examination revealed soft and cystic mass in nature. It was located in the medial aspect of the left orbit. No bruit or pulsation was noted. Everted lower eyelid with misdirected cilia were present in left eye on external examination. On Slit lamp examination micro cornea and rudimentary eye structure were noted along with poor pupillary reaction in the left eye. There was no evidence of exposure keratitis (Figure 1). Examination of visual acuity and fundus was not possible due to poor visibility of the ocular structure. Right eye examination was within normal limits. Patient had consulted various hospitals and was diagnosed to have retinoblastoma on basis of MRI. He had received a one dose of chemotherapy after an oncologist opinion. Patient was then referred to us for further management of the disease.

On detailed examination, the features were not pointing towards the diagnosis of retinoblastoma so a confirmatory B-scan was performed. B Scan revealed a cystic cavity filled with fluid along with rudimentary eye structure ruling out retinoblastoma from the differential. Based on clinical features a diagnosis of congenital cystic eyeball was made and patient was planned for cystic fluid aspiration followed by intralesional triamcinolone acetate injection. Routine haematological investigation revealed no abnormality and the chest X-ray was also normal. Cystic fluid aspiration was done under aseptic condition. About 30 cc of fluid was aspirated followed by intralesional steroid injection (Figure 1). The fluid was sent for cytological evaluation. Cytological evaluation suggested a possibility of malignancy whereas gram stain, KOH culture, and sensitivity were negative for any infective etiology.

Patient again started developing cystic swelling on the 2nd postoperative day. Oncology opinion was taken for any secondaries and systemic evaluation was done for any metastasis which were found to be negative. Patient then underwent enucleation under

aseptic conditions and the enucleated Eye was sent for histopathology. The histopathology highlighted the features of mature neuroglial tissue, fibro collagenous and vascular proliferation with adipose tissue along with areas showing glandular epithelium with mucin secretion along with mature cartilage which was suggestive of congenital mature teratoma. At the follow-up visit the socket was cleaned and dried following which an ocular prosthesis was fitted.

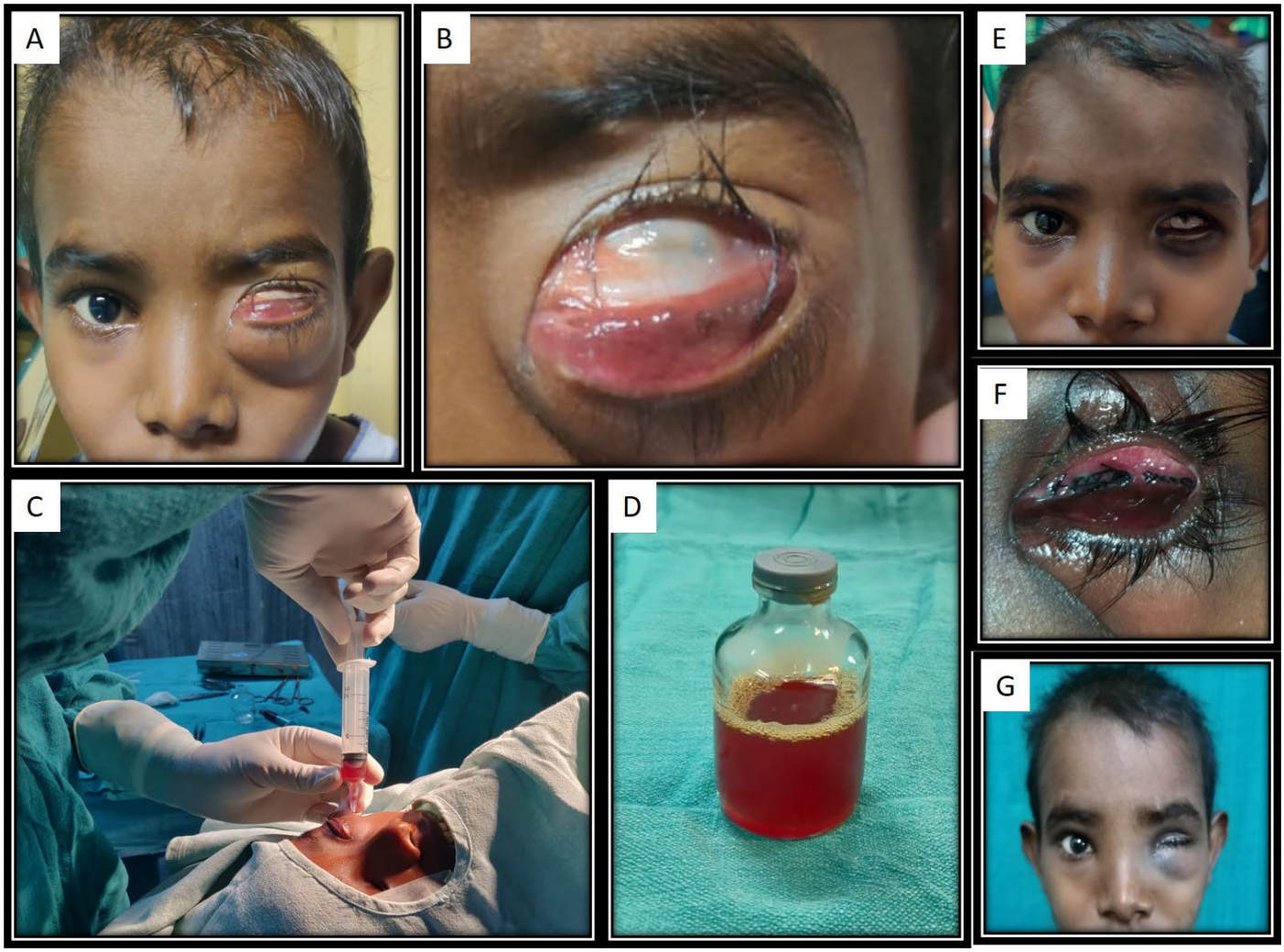

Figure 1: Features of Congenital Mature Teratoma of Eye. Large cystic swelling with misdirected cilia with microcornea in left eye (A & B). Cystic fluid aspiration being performed (C & D). Decrease in the sweeling in left eye post aspiration (E). Enucleation of left eye (F). Post-operative after Enucleation (G)

Discussion

Teratomas are masses composed of a wide variety of tissues foreign to the anatomic site from where they arise [5]. They are usually benign, and have been divided into four different groups [6]:

1. Complete orbital fetus-in-fetu (orbitopagus parasiticus).
2. Incomplete second fetus including spine.
3. True orbital teratoma with the three germ layers.
4. Dermoid tumor with only two germ cell lines.

Histopathologically, most of the teratomas are found to be comprised of two or three embryonic layers, namely ectoderm, mesoderm, and endoderm. The tumor is called a mature teratoma if the neoplastic tissue is uniformly grown and resembles the adult tissues. Tumors containing any embryonal tissue with mitotic figures are labeled as immature teratomas and these can display malignant behaviour with metastasis in future [7].

Surface Ectoderm is the predominant germ cell layer observed in orbital teratomas. Cysts filled with keratin and adnexal structures, such as hair follicles and sweat glands are also present. Neuroectodermal tissues may include primitive neural tubes, choroidal plexus, ganglia, and glial elements. The second most common germ cell layer to be found is mesoderm, which

is represented by muscle, bone, cartilage, and fat. The endoderm is the least common component of teratomas and it may produce cysts in the respiratory or gastrointestinal tissue [7]. Teratomas more common in females and it has been more frequently found in the left orbit [6]. The clinical features of these tumors include unilateral proptosis, fluctuating mass, lagophthalmos causing increased palpebral fissure, thinning of the eyelid over mass and absence of communication between the cyst and the intracranial cavity [5]. The eye may show a normal development but later may exhibit degenerative changes secondary to the displacement by the teratoma [5]. Transillumination is present in almost all part of the orbital mass due to its cystic nature [5]. Accumulation of secretions within cystic spaces causes the rapid growth of the tumor after birth. The optic nerve may be enclosed or adherent to the tumor, leading to secondary optic atrophy which may cause poor pupil reaction and decreased vision [5]. Orbital Imaging highlights, benign orbital teratomas as multiloculated, cystic swelling with an mixture of variety of tissues usually containing calcification, fat or bone.

The differential diagnosis to be ruled out are dermoid cysts, cephalocele, lymphangioma, haemangioma and epidermoid inclusion cysts [6]. The main objective in the treatment of orbital teratoma is to save the eye, to encourage orbitofacial development, maintain cosmesis, and retain some vision. Early intervention may help to preserve the globe and vision in some patients; however, preservation of the globe is not possible if no organized eye or optic nerve present [5,8].

We are keen to report this case because it is a very rare disorder and very few reports of similar cases are present in the literatures. Although in our case it was not possible to retain the eye due to the penetration of the tumor into deeper structures, so enucleation was the preferred choice of treatment. The reconstruction process was successful in spite of the great tissue deformity with a very good cosmetic result.

Conflict of Interest. No conflicting relationship exists.

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