# Management of a Congenital Cyst of the Orbit in a Five Year Old

Hadia Yousuf[1], Uzma Fasih[2], Erum Shahid[3], Arshad Shaikh[4], Asad Jafri[5]

## Abstract

We report here the case of a 5 year old female child who presented at Eye department of Abbasi Shaheed Hospital with gradually increasing proptosis of left eye ball since birth. There was no light perception in the eyeball. Examination revealed a cystic swelling in inferolateral part of the orbit causing superomedial displacement of the orbit with restriction of extraocular movements. In addition there was microcornea, miccrophthamos and coloboma of iris extending posteriorly to the optic nerve. Right eye showed microphthalmos, microcornea, nystagmus on attention and a reacting pupil with visual acuity of finger counting at two feet. CT scan showed a large smooth walled hypodense lesion arising from inferolateral aspect of the orbit displacing the eye ball superomedially along with remodeling of adjacent bones.The cyst was successfully removed through subcilliary incision under general anaesthesia. Continuation of the cyst with optic nerve was seen peropeatively. Histopathology revealed a benign cyst with lining of single layer of epithelium and wall composed of aggregates of lymphocytes congested blood vessels and skeletal muscles. Findings were consistent with colobomatous cyst of the orbit.

**Keywords:** Orbit, congenital cyst, microphthalmos.

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## Introduction

A large variety of cystic lesions may involve the orbit. They may be subdivided into developmental anomalies and acquired lesions. These include dermoid cysts, colobomatous cysts, certain vascular lesions with major cystic components such as lymphangiomas, lacrimal duct cysts, haematic cysts, epithelial and appendage cysts, parasitic cysts, cephaloceles and mucoceles. Rare lesions such as optic nerve cysts and enterogenous cysts of the orbit have also been described in the literature[1-4]. Dermoid cysts represent the most common congenital lesion of the orbit and account for one third of all childhood orbital tumors[5]. The superior temporal quadrant at the frontozygomatic suture is the most common location, followed by the nasal aspect of upper orbit at the frontoethmoidal suture. These arise as developmental sequestration of the ectoderm within the suture lines or diploe of the orbital bones. This explains the close relationship of these lesions with the sutures of the orbit. Orbital dermoids may be subdivided into superficial or deep lesions. The superficial dermoids present in infancy as painless subcutaneous nodules near the orbital rim and are not associated with proptosis; deep lesions are usually larger, seen in adults, most commonly arise behind the orbital rim and may show proptosis or displacement of the globe[6].

A coloboma is a congenital or acquired notch, gap or fissure in which a portion normal tissue is absent. Any part of the eye may be affected, how-

[1] Jinnah Sindh Medical University
[2-5] Eye Department, Abbasi Shaheed Shaheed Hospital and Karachi Medical and Dental College, Karachi.

**Correspondence: Dr. Uzma Fasih**
**Eye Department**
**Abbasi Shaheed Shaheed Hospital and**
**Karachi Medical & Dental College**
**Email: yousufuzma@hotmail.com**
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ever, typically the cleft appears in the inferonasal quadrant of the globe[2]. The optic nerve head is also commonly involved resulting in central excavation of the disc. More than 60% of cases are bilateral[6]. A coloboma may be an isolated finding or a part of a complex cleft in the globe[7]. A colobomatous cyst is defined as a neuroectoderm-lined mass that protrudes through a coloboma in the wall of a microphthalmic eye[8]. It is usually diagnosed at birth and can affect one or both globes. Colobomatous cyst of the orbit is a rare congenital cystic malformation associated with ocular maldevelopment. Usually, the cyst is associated with a microphthalmic globe[9]. On imaging, the affected eye may be normal sized or microphthalmi. The tunnel-like connection between the eye and the cyst is often very thin and difficult to image. CT scan demonstrates the anatomic relationship of the cyst to the globe prior to surgical intervention as well as any associated developmental anomalies of the brain. We report this case here to elaborate the presentation; evaluation and management of congenital orbital cyst as these type of cysts are rare entities.

**Case Report**

A five-year-old female child presented at Eye department of Abbasi Shaheed Hospital Karachi in October 2015 with gradually increasing proptosis of left eyeball since birth. Regarding the visual acuity there was no light perception. Examination revealed a cystic swelling in the inferolateral part of the orbit anteriorly causing superior and medial displacement ofleft eyeball. Ocular examination of left eye revealed the restriction of extra ocular movement in all gazes but more in lateral and inferior gaze. There was microopthalmos and micro cornea, along with coloboma of iris extending posteriorly to choroid and optic nerve. Right eye showed micropthalmos microcornea, nystagmus on attention and a reacting reacting pupil with visual aquity of finger counting at two feet Fig. 1. There was no significant finding on general physical examination. B scan showed multiloculated cystic lesion measuring approx. 3.5 X 2.7 cm with low level seen at the lateral and inferior aspect of the eye ball. Lens appears normal in both eyes. Minimal vitrious degeneration was present bilaterally. A CT Scan with non-ionic contrast was ordered which showed a large smooth walled hypodense lesion arising from the inferolateral aspect of left orbit. It was abutting the sclera and pushing the eyeball medially and superiorly. There was no calcification, fatty density or abnormal contrast enhancement was seen. There was remodeling of adjacent bones with enlargement of bony orbit seen. The mass has remodeled the left maxillary sinus. The adjacent ethmoid sinus was normal. Right orbit showed no abnormality. Investigations for general anaesthesia (GA) were done and surgery was planned under GA. The cyst was successfully removed enblock through subcilliary incision Fig. 2. Extension of cyst up to optic nerve was found peroperatively. Histopathology showed cyst wall lined by single layer of epithelium. The wall composed of lymphocytes aggregate congested blood vessels and skeletal muscles. A diagnosis of benign cystic lesion was made which could be a colobomatous congenital cyst. Permission of the parents was taken for the pictures to be published.

The patient developed an inability to close the left eye postoperatively so a tatsorrhaphy was done postoperatively to prevent exposure keratitis Fig. 3.

**Discussion**

A majority of the cases of colobomatous cyst are associated with microphthalmos. With no specific preponderance for any particular gender. The condition is usually unilateral[8]. The etiology of a colobomatous cyst is not exactly known but it is presumed to occur due to improper fusion of the embryonic fissure between the 7-14 mm stage of fetal development. This results in abnormal ectasia of the sclera, which expands into the adjacent orbit[10].

The presentation of microphthalmos with a cyst can be seen as a protruding mass in the inferior orbit associated with a malformed microphthalmic eye.The cyst may be so small that it

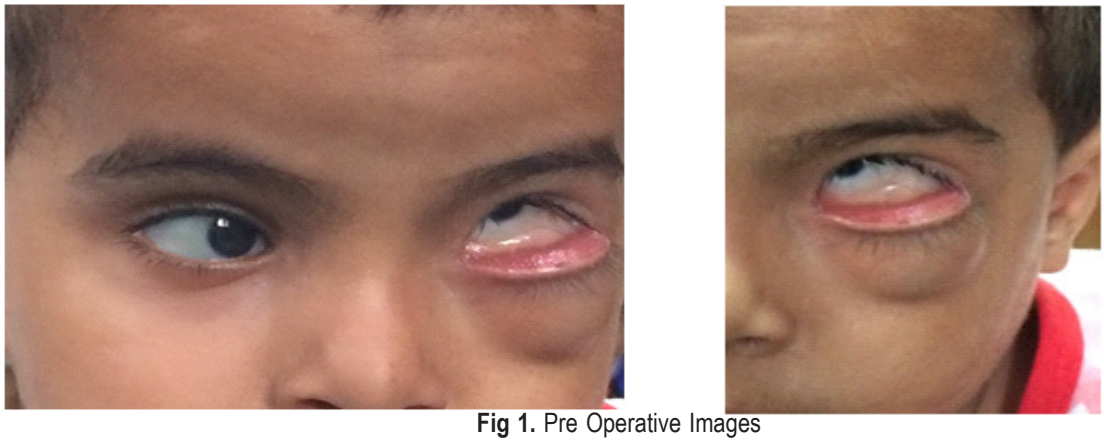

**Fig 1.** Pre Operative Images

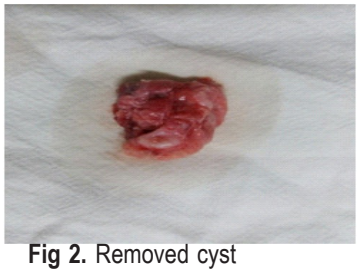

**Fig 2.** Removed cyst

**Fig 3.** Post Operative Images

cannot be detected clinically or it may be so large that it obscures the globe[11]. These eyes usually have a poor visual outcome. Foxman and Cameron reported that bilateral microphthalmos with colobomatous cyst may be associated with major systemic abnormalities (central nervous system, renal, or cardiovascular), whereas unilateral involvement is usually associated with minor abnormalities[12]. Our case was a unilateral presentation fortunately with no systemic abnormality.

Imaging is of primary importance in the evaluation of cystic lesions of the orbit as ithelps in revealing any communication between the cyst and the globe. This information is useful for planning the management of these lesions. Management of these cysts depends on the age of the patient, the size of the cyst, and the presence of communication between the globe and the cyst, and the visual prognosis. Surgical management varies from simple aspiration of the cyst, enucleation of the microphthalmic eye along with the cyst, and excision of the cyst with preservation of the globe[13]. We managed our case by aspiration of cyst and its excision through subcilliary incision. Unfortunately the patient developed inability to close the eyelid postoperatively so tarsorrhaphy was done to prevent exposure.

Once the cyst has been removed, if the patient is anophthalmic and there is no significant microphthalmic remnant, an orbital implant can be inserted to maintain the orbital volume and aid cosmesis and orbital growth. Both silicone and hydroxyapatite have been used as orbital implants[14-15].

### Conflict of interest

Authors have no conflict of interests and no grant/ funding from any organization.

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