CASE REPORT

Bilateral Anophthalmia with Lymphangioma of Bilateral Orbit, A Rare Congenital Ocular Presentation: A Case Report

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ABSTRACT
In the present case report we describe a rare case of a 3-year-old male patient with a history of absence of both eyeball since birth now presenting with tender cystic swelling of right lower preseptal and orbital area with discolouration of the skin. There is history of intermittent discharge since 1 year. This case report highlights the step wise approach to diagnose a case of bilateral anophthalmia presenting with lymphangioma of bilateral orbit and its management by percutaneous sclerotherapy with injection bleomycin leading to promising results.

Keywords – Anophthalmia, lymphangioma, percutaneous, sclerotherapy, bleomycin, case report

Introduction

Anophthalmos (anophthalmia) refers to the complete absence of any visible globe structure, though a microphthalmic remnant or cyst may be present [1]

Orbital lymphatic-venous malformation (previously called lymphangioma) is an uncommon unencapsulated vascular malformation of the lymphatic system generally present in childhood [2]. These benign cystic lesions are characterized by abnormal endothelial-lined channels [3] and represent 1%-4% of all orbital masses [4,5].

Clinically, orbital lymphangiomas can be asymptomatic. Some patients with orbital lymphangioma may develop proptosis, either slowly as the mass invades the orbit or suddenly during hemorrhage of a lesion [3,6]Several methods have been used to treat orbital lymphangioma, including systemic corticosteroids, injection of a sclerosant, and surgical excision, but currently, there are no definitive curative treatments.

Here we discuss a rare case of bilateral anophthalmos presenting with biorbitallymphangioma(right larger than left) and its treatment with injection bleomycin in the right sided lesion

Case Presentation:

A 3-year-old male presented with absence of both eyeballs along with tender cystic mass in the right orbit and preseptal region associated with skin discolouration and intermittent discharge over a period of 1 year. The mother of the patient complained that there was absence of both the eyeball since birth and that the patient cannot talk or listen or walk as compared to his peers of the same age group. The child was a term baby born via normal vaginal delivery and birth weight of 2.5 kg. There was history of neonatal ICU admission for 8 days. There was no history of ocular trauma or surgery.

Upon examination, lids of right eye was swollen and ecchymosed with presence of a cystic mass of size 3x4x3.5 cubic cm was present in the hollow of right orbit. The left preseptal region also had similar smaller swelling

Figure 1- presentation at admission

ON further workup-

Fine Needle Aspiration Cytology (FNAC) right orbit region swelling revealed smears consisting of mostly RBC and other hematopoetic cells (neutrophil, lymphocytes and eosinophils only). No other cellular elements identified.

Ultrasonography (USG) bilateral orbit revealed both orbital fossa as empty and replaced by multiloculated cyst structures and septations in the right orbital fossa of size 3.8 x3.8 square cm. Similar smaller size lesion is seen in the left orbital cavity.

Contrast Enhanced Computed Tomography (CECT) orbit revealed a multiloculated cystic lesion of size approximately 3x4 cm square is noted . There was enhancement of the septae and the walls of lesion. Similar lesion of smaller size was seen in the left orbital cavity.

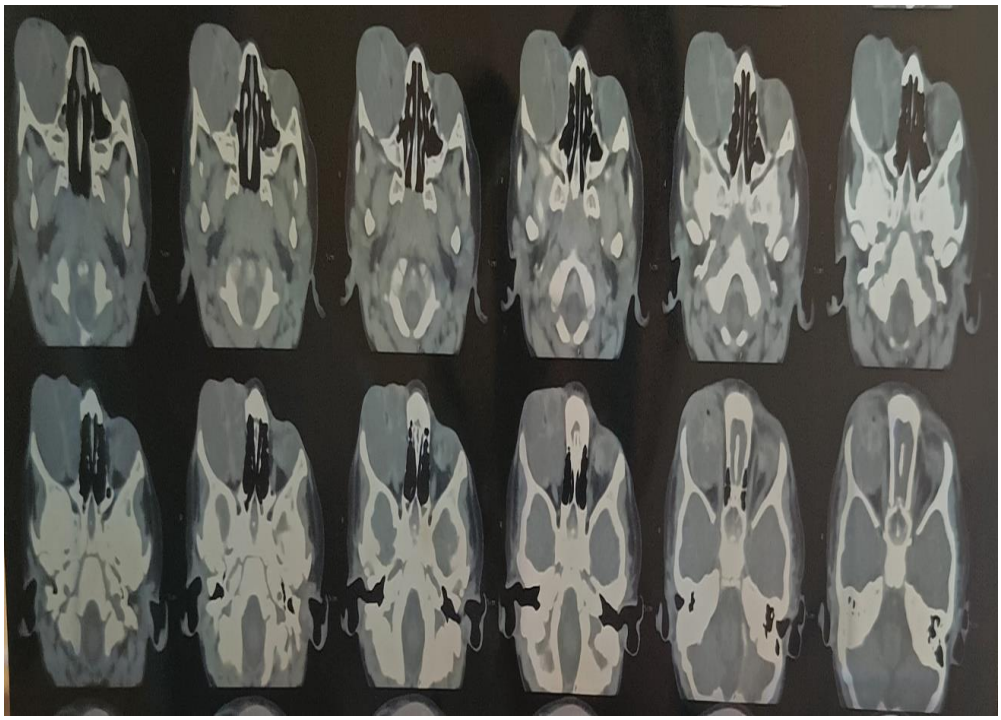
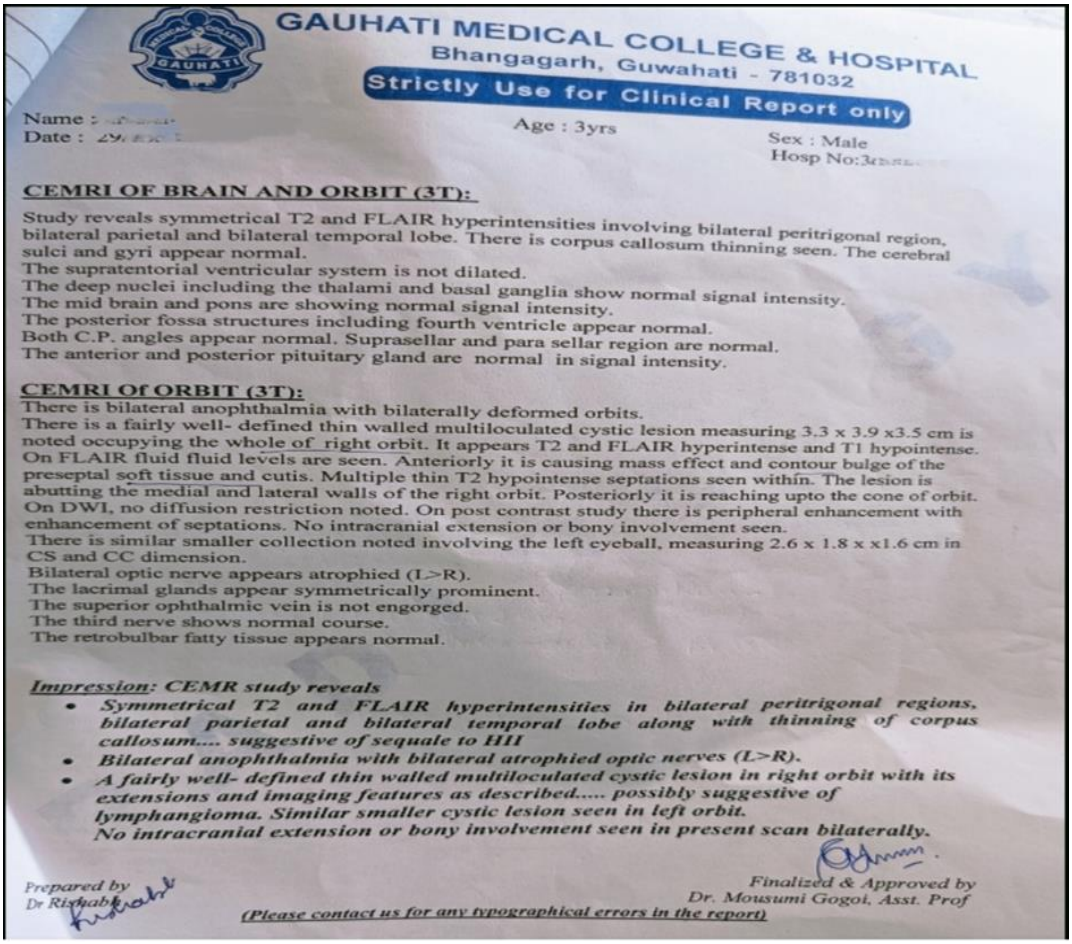


Figure 2- CECT orbit plate of the patient revealing multiloculated cyst in the right orbit.

Magnetic Resonance Imaging (MRI) reveals bilateral anophthalmos with bilateral atrophied optic nerve (Lt >RT) and suggests lymphangioma right orbit with similar smaller similar cystic lesion in the left orbit



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Name :
Date :
Age : 3yrs
Sex : Male
Hosp No:

CEMRI OF BRAIN AND ORBIT (3T):

Study reveals symmetrical T2 and FLAIR hyperintensities involving bilateral peritrigonal region, bilateral parietal and bilateral temporal lobe. There is corpus callosum thinning seen. The cerebral sulci and gyri appear normal.
The supratentorial ventricular system is not dilated.
The deep nuclei including the thalami and basal ganglia show normal signal intensity.
The mid brain and pons are showing normal signal intensity.
The posterior fossa structures including fourth ventricle appear normal.
Both C.P. angles appear normal. Suprasellar and para sellar region are normal.
The anterior and posterior pituitary gland are normal in signal intensity.

CEMRI Of ORBIT (3T):

There is bilateral anophthalmia with bilaterally deformed orbits.
There is a fairly well- defined thin walled multiloculated cystic lesion measuring 3.3 x 3.9 x3.5 cm is noted occupying the whole of right orbit. It appears T2 and FLAIR hyperintense and T1 hypointense. On FLAIR fluid fluid levels are seen. Anteriorly it is causing mass effect and contour bulge of the preseptal soft tissue and cutis. Multiple thin T2 hypointense septations seen within. The lesion is abutting the medial and lateral walls of the right orbit. Posteriorly it is reaching upto the cone of orbit. On DWI, no diffusion restriction noted. On post contrast study there is peripheral enhancement with enhancement of septations. No intracranial extension or bony involvement seen.
There is similar smaller collection noted involving the left eyeball, measuring 2.6 x 1.8 x x1.6 cm in CS and CC dimension.
Bilateral optic nerve appears atrophied (L>R).
The lacrimal glands appear symmetrically prominent.
The superior ophthalmic vein is not engorged.
The third nerve shows normal course.
The retrobulbar fatty tissue appears normal.

***Impression:* CEMR study reveals**

- ***Symmetrical T2 and FLAIR hyperintensities in bilateral peritrigonal regions, bilateral parietal and bilateral temporal lobe along with thinning of corpus callosum.... suggestive of sequale to HII***
- ***Bilateral anophthalmia with bilateral atrophied optic nerves (L>R).***
- ***A fairly well- defined thin walled multiloculated cystic lesion in right orbit with its extensions and imaging features as described..... possibly suggestive of lymphangioma. Similar smaller cystic lesion seen in left orbit.***
  No intracranial extension or bony involvement seen in present scan bilaterally.

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(Please contact us for any typographical errors in the report)

Figure 3- MRI report establishing anophthalmia both eye with lymphangioma Right orbit

Management

Since the right sided lesion was larger and symptomatic it was decided to do staged bleomycin injection. The left orbital lesion was to be treated at a later date.

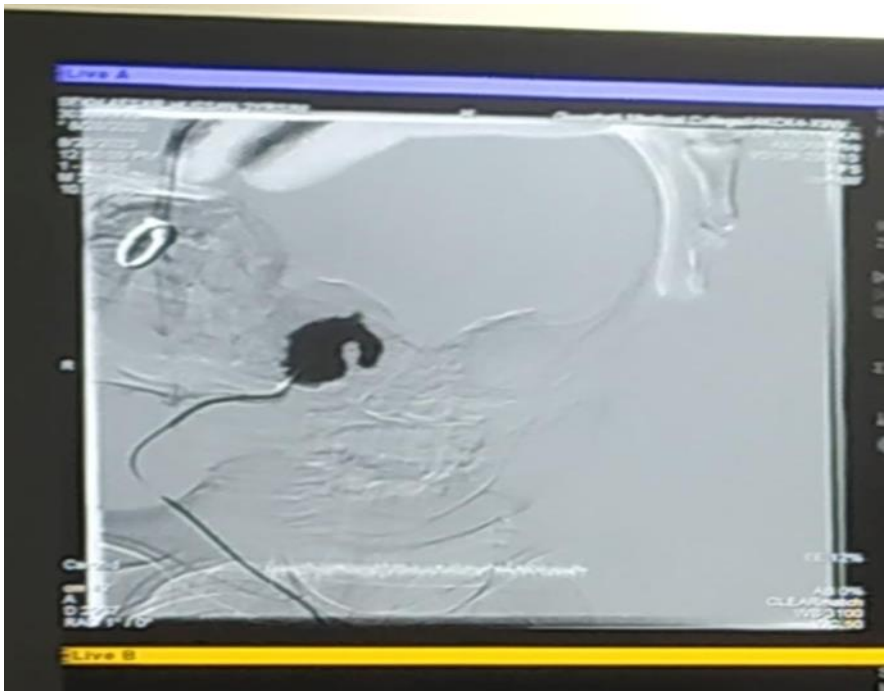

Figure 4 – Digital Subtraction Angiography (DSA) image acquired before bleomycin injection to see for direct communication with orbital vein during the sclerosant therapy procedure

The patient underwent percutaneous sclerotherapy via scalp vein with bleomycin injection under conscious sedation. 10ml of fluid was first aspirated and 9IU of bleomycin injected in the orbital lymphangioma followed by 2ml of sodium tetradecyl sulphate foam. Compression bandage was applied.

As in the immediate post operative period, patient was likely to develop swelling Injection Dexamethasone – 1cc iv twice daily from 1 day before the treatment upto 5 days followed by 1cc i.v once daily for next 5 days was given

Injection Ceftriaxzone -0.5 gm I.v once daily for 5 days along with t Topical Moxifloxacin eye drop and eye ointment for 6 times and 3 times Rt eye for 2 weeks was also advised

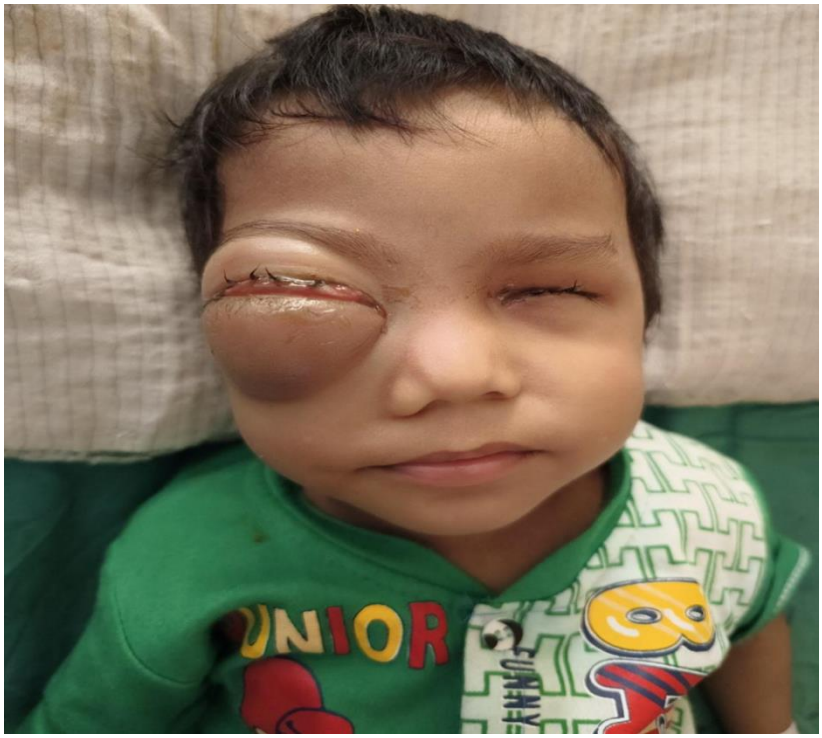

Figure 5- Image of the child immediately following sclerosant therapy

Figure 6- Image of the child before discharge

The child is asked to review after 3 months and further line of management will be decided after that visit.

Results:
The staged approach to diagnose and management of bilateral anophthalmos with right orbital lymphangioma wielded favourable results. The patient experienced a significant reduction in the size of the right orbital lymphangioma mass and also reduction in the pain after the treatment with the bleomycinsclerotherapy.

Discussion:
Approximately 25% of orbital lymphangioma cases involve the orbit (7).
Orbital lymphangiomas are currently classified as Type 1 (low flow and no vascular system connection) orbital vascular malformations by the International Orbital Society. Any lesions that are found to have any vascular system connection fall under Type 2 (8)
Imaging with MRI can be useful to plan management. Although computed tomography allows assessment of the bony orbit and surrounding facial bones, this is not useful for planning sclerotherapy. Ultrasound scans are useful for assessing the orbit post-treatment.
Sclerosants have been used for treating lymphatic malformations for several years. Agents that have been previously reported as being used for orbital lymphangioma include OK-432 (Picibanil; Chugai Pharmaceutical Co Ltd), sodium tetradecylsulfate (multiple manufacturers), 5% sodium morrhuate, and bleomycin A5. (9-13). Bleomycin is the drug of choice for lymphangioma especially in locations like orbit as it causes lesser degree of post injection swelling.

Conclusion:
Management of orbital lymphangioma continues to require a multi-disciplinary approach, and early involvement of other specialities when needed can prevent delays in arranging imaging or treatment. It is not unusual nowadays for treatment to comprise of both surgical and non-surgical aspects in order to remove/shrink as much of the lesion as possible while minimising visual and functional loss. It is important to remain in frequent communication with the patient and/or their parents, as well as colleagues, to ensure a consistent message and plan of action. What is clear is that percutaneous sclerotherapy is now an established management option for these lesions, with good results (and minimal complications) in appropriately chosen patients (14)

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Conflict of interest: Nil
Financial interest: Nil

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