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A Rare case scenario of extranasal congenital glial heterotopia

Dr. Hirva patel , Dr. Rabab ginwala, Dr. Sapan shah , Dr. Nishtha patel ,

Resident doctor , AMC MET Medical college , Ahemdabad
M.S ophthalmologist, Ahmedabad
Oculoplastic surgeon , Ahmedabad
Senior Resident , AMC MET Medical college, Ahemdabad

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ABSTRACT::
Purpose:Purpose is to represent a rare case of glial heterotopia in a eye which was not originating from the medial bone suggestive of extranasal origin.
Observation:A 3 months old child having microphthalmia with cystic lesion for which excisional biopsy was done with histopathology report suggestive of glial tissue.
Conclusion:So it is a rare case of glial heterotopia in which glial tissue proliferate from midline and not from the nasal bone .So, like in this case early diagnosis with MRI would be more helpful in cosmesis.
Keywords: Ectopic brain tissue, Orbit, Microphthalmia

I. INTRODUCTION:

Microphthalmia is characterized by small but recognizable eye with eye elements such as choroid, lens and retina.(6) In contrast to this, anophthalmia is characterized by the complete absence of the eye due to the lack of development or arrest of differentiation of the optic vesicles in early stage of development. Glial heterotopia also known as Nasal glioma represents collections of normal glial tissue at abnormal position distant to the central nervous system with no intracranial connectivity.The most common reported site is in and around the nose. Although benign, they can cause significant local damage and cosmetic deformity by compressing orbital walls. Gliomas form an uncompressible mass that does not increase in size on the Valsalva testing and does not transilluminate. Confirmatory diagnosis of glial heterotopia can be done with the help of imaging and histopathological findings supported by immunohistochemical presence of glial fibrillary acidicprotein and S100 protein. Surgical excision is the mainstay of treatment for gliomas, but they must be differentiated from encephalocele and other midline nasal masses before the surgery is scheduled.(7)

II. CASE REPORT:

A 3-month old female child presented with the chief complaint of pinkish swelling in her left eye since 17 days,as informed by her parents. Past medical history suggestive of black swelling present in left orbit instead of normal eye at the time of birth. At the age of 1.5 months patient went to a general ophthalmologist for the same and sonography was done. On sonography of left eye, partially deformed cystic shaped swelling was seen in left orbit anteriorly. Posterior to swelling a polypoidal echogenic structure was seen suggestive of eyeball but chamber differentiation and lens visualisation could not be done. Neoplastic etiology was suspected. Patient was referred to ocular oncologist. At the age of 3 months patient was brought to our centre with keratinization of swelling.On examination large pedunculated pinkish swelling protruding outside the eyelid was present in left orbit instead of eye (figure 1). Patient sent for MRI of orbit with brain (figure 2). MRI report suggestive of cystic swelling present in left orbit with echogenic structure behind swelling, ((?) Microphthalmic eye) with no differentiation of eye structures. There was no intracranial communication or extension. Anterior orbitotomy was performed and removal of the cyst en mass was done. After removal of the cyst (figure 3), it was sent for histopathological examination and eyeball was found to be atrophic. Histo-path examination revealed glial heterotopia (figure 4), i.e. brain tissue at ectopic place. Based on these findings, the diagnosis was right eye within normal limits with left eye having microphthalmos with intraorbital cyst.

III. DISCUSSION:

Microphthalmia is considered to be the second most common congenital malformation of

the eye after congenital cataract. The detection of ectopic glial tissue can be done in early neonatal period. It typically presents as a protruding mass in the orbit associated with microphthalmic eye. The globe may be completely surrounded by the cyst in some cases, while others may present with very rudimentary displaced microphthalmic eye, thus creating difficulty in identifying the eye clinically as in our case. In this type of cases orbital ultrasound with MRI scan is of great importance in identifying orbital cysts. Although it is clearly a defined entity, sometimes it becomes difficult to differentiate it from some lesions such as congenital cystic eye, meningocele, arachnoid cyst, primary optic nerve sheath cysts, and teratomas of the orbit. Lieb et al. has mentioned the importance of imaging techniques in ruling out these differentials [3]. In congenital cyst with microphthalmia, the cyst is located centrally or slightly upward in the orbit and histologically it lacks normal ocular structures and shows the presence of the cyst lined with neuroglial tissue. The treatment strategy for patients with orbital cyst depends on several factors such as visual potential, age at presentation, and volume of the orbital content. Chaudhry et al. have proposed the treatment protocol for congenital orbital cyst. In case of mild microphthalmia, if the cyst is small, cyst aspiration and observation is recommended, whereas if it is large affecting cosmesis, excision of the cyst should be performed like done in our case. In case of severe microphthalmia, both the cyst and globe should be excised with replacement of volume. Repeated follow up should be kept in consideration for early detection of recurrence or neoplastic transformation.

IV. CONCLUSION:

Here , we report a rare case of congenital microphthalmia with glial heterotopia of extranasal origin. Several studies are still required to understand the spectrum of this complex condition, which is considered as one of the causes for childhood blindness, more commonly unilateral. Advanced imaging technology specially MRI can be helpful in early diagnosis of the lesion which in turn provide great help in counseling the parents and provide appropriate treatment options in early neonatal life. Even cosmetic care should be taken care of immediate after cyst removal to prevent socket contraction and disfigurement of eye.

Conflict of interest: -
No conflict of interest

Consent :

Informed consent was obtained from the parents for present this case

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Figures:

Figure 1

Figure 2

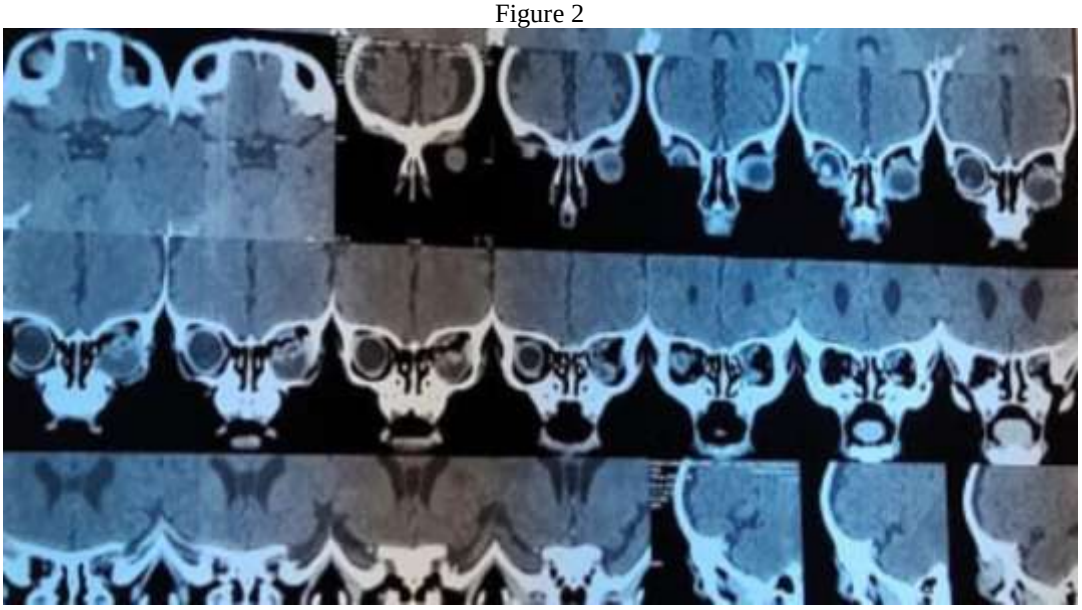

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Figure 3

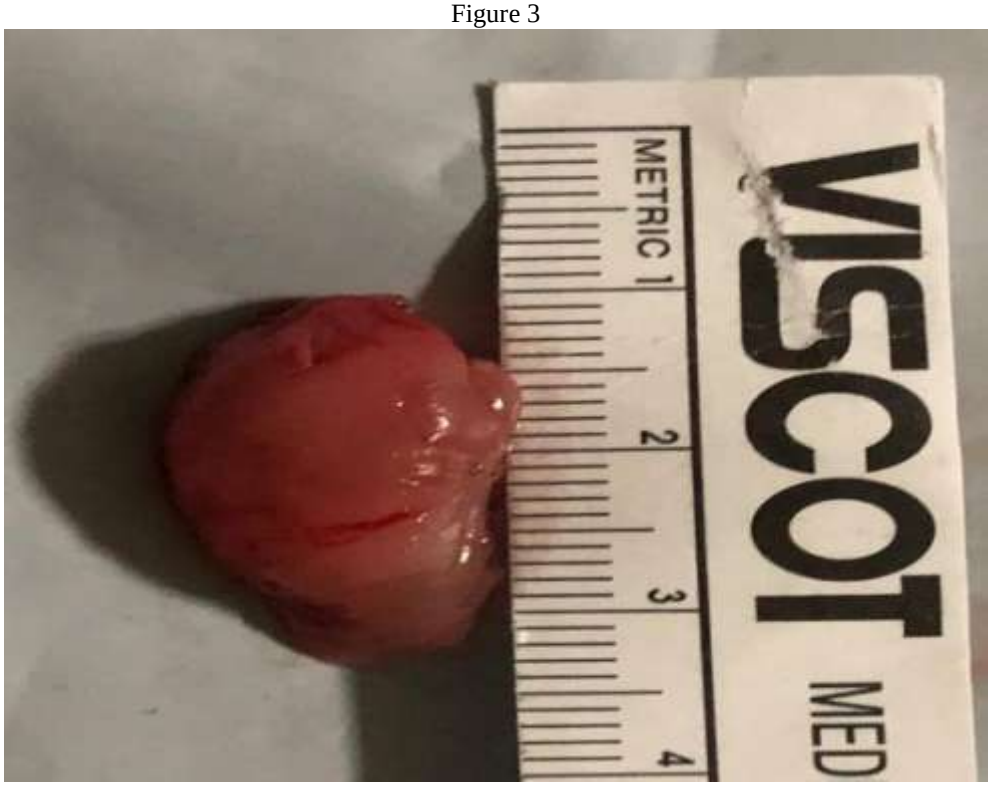
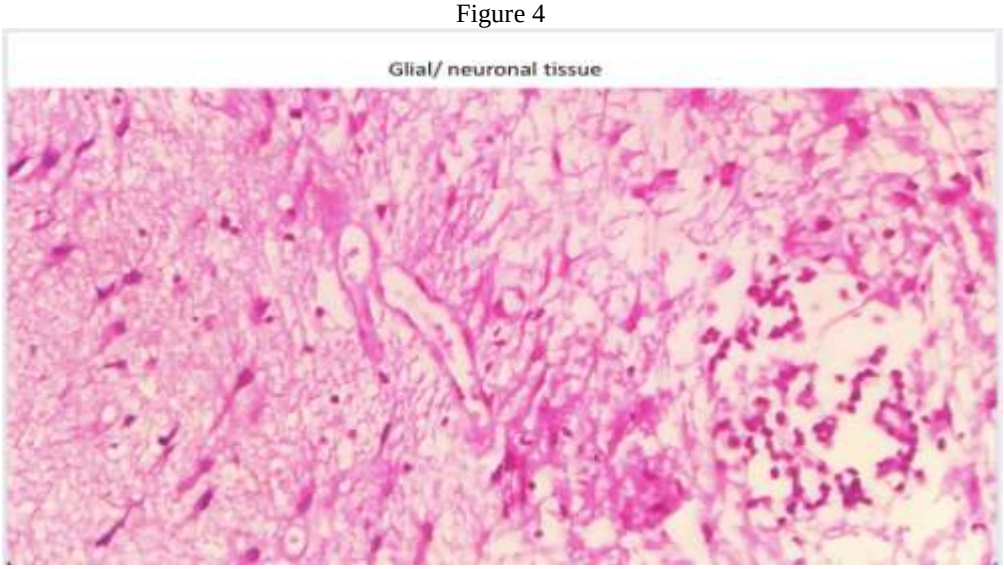


Figure 4



Caption:
Figure 1: Preoperative view of left eye.
Figure 2: MRI showing cyst with Microphthalmic eye.
Figure 3: Cyst which was removed from left orbit.
Figure 4: High power view of glial tissue 400x.