
CASE REPORT

Bilateral Orbital Meningocele Without Frontoethmoidal Meningocephalocele: A Rare Presentation

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ABSTRACT

Introduction: Bilateral orbital meningocele is a rare congenital abnormality. In our review, no case was reported from Southeast Asia which has reported bilateral orbital meningocele with frontoethmoidal meningocephalocele. Frontoethmoidal meningocephalocele has many extracranial manifestations out of which orbital meningocele are one of them. In this study, we are reporting a case of a 4 years old male child presenting with bilateral exophthalmos and gradual loss of vision bilaterally. He underwent CT and MRI that showed bilateral intraorbital, extraconal mass lesions displacing the orbit. Cribriform plate was low lying with herniation of brain parenchyma in frontal region anteriorly. These findings were later proved to be similar post operatively.

Keywords: Orbital meningoencephalocele, cephalocele, meningocele.

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INTRODUCTION

Orbital meningocele, herniation of meninges along with cerebrospinal fluid into the orbit, is a very rare congenital abnormality. The herniation usually occurs by a defect in the bony orbit or, more rarely, can occur in the openings naturally present such as sphenoid.¹ It may be associated with cephalocele can be further classified by its location as either anterior or posterior.

A review of 250 orbital biopsies performed in children has reported only one cephalocele.² A defect between frontal and lacrimal bones may lead to anterior ethmoidal cephalocele. This usually appears as a smooth and fluctuant swelling on the side of the nose adjacent to the medial canthus. This may be bilateral in some cases. Characteristic rhythmic pulsations make cephalocele presence appreciable. CT may demonstrate bony defect and MRI demonstrates the characteristics of the meningocele.²⁻³ Large lesions may require surgical removal.²⁻³

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CASE REPORT

A 4-years-old male child presented with a history of bilateral periorbital swelling that was present since birth but gradually increased in size. He had gradual loss of visual acuity in both eyes. On physical examination soft, fluctuant swelling was identified. There was also bilateral exophthalmos. He underwent both CT and MRI brain. CT brain demonstrated low lying cribriform plate of ethmoid with herniation of brain parenchyma downwards in frontal region anteriorly. Another bony defect in naso-orbital region between the nasal and lacrimal bone was identified bilaterally with herniation of meninges through them. Large lobulated, multiseptated cystic, intraorbital, extraconal lesions were also identified involving bilateral orbits medially causing remodeling of the orbital bones without erosion or destruction. These were significantly displacing the globe with optic nerves and extraocular muscles superolaterally. MRI demonstrated these masses as multiseptated cystic lesions with T1 and FLAIR hypointensity and T2 hyperintensity causing displacement of globe laterally and compression of optic nerve. Minimal brain tissue was protruding through the bony defect bilaterally suggesting possibility of Orbital Meningoencephalocele. Coronal images showed lower down cribriform plate of ethmoid with cystic lesions and herniation of the

meninges and brain tissue through bony defect in the region of lacrimal and nasal bone bilaterally. He underwent decompression surgery with craniofacial repair and reconstruction of medial canthus. Surgical findings were consistent with meningoencephalocele. On postoperative follow up examination he underwent MRI and CT again. CT showed post-surgical changes with bone grafting along medial wall of orbit. Both CT and MRI showed interval reduction in size and mass effect of intraorbital cystic masses. Post-operative course was unremarkable with event free recovery.

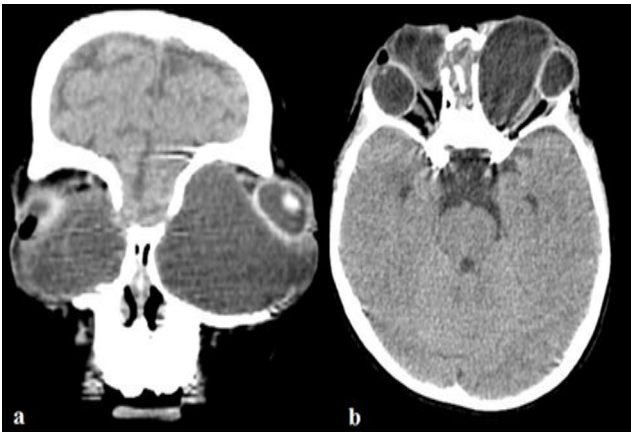


Figure 1: CT scan of 4 years old male. (a) Coronal section showing low line cribriform plate. (b) Axial section showing defect in nasolacrimal bilaterally with multiseptated cystic intraorbital extraconal lesions.

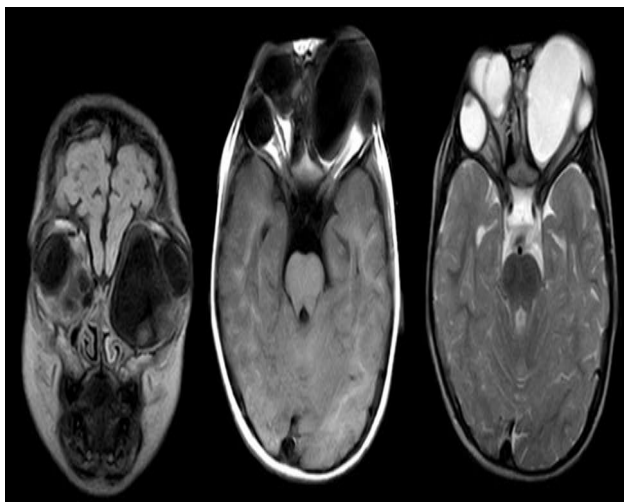


Figure 2: MRI brain (a) Flair coronal (b) T1 Axial (c) T2 Axial showing lobulated and septated cystic extraconal lesion appearing hyperintense on T2 and hypointense on T1 and flair images

DISCUSSION

Orbital meningocele is a rare cystic lesion of childhood usually found in association with fronto-ethmoidal meningoencephalocele, which is a neural tube defect with herniation of brain parenchyma and the meninges

through a bone defect at the skull base at the junction of the ethmoid and frontal bones. Nasoorbital meningoencephalocele is a rare type of frontoethmoidal meningoencephalocele.³

This defect is reported as 1 in 3500 live births in South East Asia.⁴ However in our case no bony defect was evident at skull base and only cribriform plate was abnormally low leading to brain herniation in frontal region, making it a rare finding. Influence of environmental factors and deficiencies of nutrients such as folic acid and various drugs with teratogenic effects have been suspected as its possible causes. This is a frequent cause of facial disfigurement, nasal function impairment, disturbances in vision or any CNS infection. Infancy period is the optimal time for definitive correction to prevent further growth disturbances and serious complications⁵. Our case was unusual for a reason that bilateral orbital meningoceles that present without definite evidence of frontoethmoidal meningoencephalocele. This has not been reported from Asian population previously. In cases of large orbital meningoceles prompt management measures should be undertaken to prevent vision loss.

CONCLUSION

Bilateral intraorbital meningocele without definite evidence of frontoethmoidal meningoencephalocele is a rare presentation that should be treated promptly to prevent vision loss and other complications.

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