CASE REPORT

Meningoencephalocelewith Coexisting Large Arachnoid Cyst: Case Report of ayoung adult of South Asian origin

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SUMMARY

Meningoencephalocele or simply known asencephalocelesare rare congenital anomaly in which brain tissue herniates out through a defect in the cranium. They occur one in every 4000 live births. Depending on the site of defect, major types are posterior oroccipital type (85% of cases) , anterior or nasofrontal type (15% of cases) and basal (10% of cases)[1]. Anterior encephaoceles are commoner in South and Southeast Asian populatuon[2,3]. Most cases present in infancy[4].The associated findings on imaging are hydrocephalus (15%), corpus callosum agenesis (7%), arachnoid cyst (3%), porencephalic cyst (3%) and single ventricle (3%)[5]. Our case is unique as the presentation is in 25 years of age andthere ispresence of a associatedlarge arachnoid cystin posterior and middle cranial fossa.

Keywords: Encephaocele, arachnoid cyst, meningoencephalocele

CASE REPORT

We present a case of a 25 years old male who presented to the Radiology departmentof Jinnah hospital, Lahore withavisible swellingover thebridge and the root of the nose. The swelling was present since birth and has remained stable in size since then. History and review of systems were unremarkable except history of occasional headaches.On examination there was 5x7cmsoftswellingover the bridge of nosewas noted. It was neither pulsatile, warm or tender to touch. There wasmoderate degree of hypertelorism.Neurlogicalexamination including power,tone, reflexes, cerebellar function, sensory system andspeechwere unremarkable. Vision was normalbilateraly.Ophthalmoscopic examination revealed mild papilledema. Cross sectional imaging including CT and MRI were conducted and revealeda1.9 cm sized defect in frontoethmoidal region.Through this defect frontal lobe and meninges were herniating out to form a swelling over the bridge of nose in intercanthal region. This herniated tissue was causing mass affect andlateral deviation of both eyeballs however no evidence of optic disc edema. Posteriorly a huge cerebrospinal fluid (CSF) intensity lesion was appreciated in posterior fossa. This lesion was extending upwards from the posterior fossa through the tentorium cerebelli extending into the right sided middle cranial fossa apical wall. The lesion was causing severe mass effect on the temporal lobe, displacing it laterally, parietal lobe was displaced superiorly and the right lateral ventricle was also displaced and compressed while the contralateral lateral ventricle showed mild prominence. The cerebellum was displaced inferiorly by the lesion with resultant severe tonsillar herniation of 1.9cm.The lesion showed suppression of signals on FLAIR sequences and absence of any restricted diffusion. There was no underlying gliosis in frontal lobe. Thiscase was diagnosed asfrontoethmoidalencephalocele with an associatedarachnoid cyst. The patient was referred to neurosurgery for repair of encephalocele with underlying bony defect closure as well as aspiration of arachnoid cyst.

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DISCUSSION

Encephaloceles are rare congenital anomalies thought to be due to incomplete closure of rostral end of neural tube. The resultant bony defect along the midline of cranial vault or base of skull results in herniation brain and meninges through it[6]. They occur one in every 4000 live births. There is no recognized gender predilection and most cases are sporadic in origin. The major types are posterior or occipital type (85% of cases) , anterior or nasofrontal type (15% of cases)and basal (10% of cases)[1].Other varieties include lateral parietal and orbital encephaloceles. Anterior encephalocele is characterized into various types among which frontoethmoid type is the commonest (nasofrontal, nasoethmoid and naso orbital), followed by transethmoidal, trans sellar or trans sphenoidal,

anterior fontanelleand interfrontal types.Frontoethmoidalencephaloceles are also known as sincipitalencephaloceles. These encephaloceles are more common in South and Southeast Asian populations [9] probable thought to be due to underlying folic acid deficiency in the population. Nasofrontal types are found projecting along the nasal bridge between the nasofrontal sutures into the glabella.Nasoethmoidencephaloceles project under the nasal bones and above the nasal septumand Naso orbital types project along the medial orbit at the level of the frontal process of the maxilla and the ethmoid-lacrimal bone junction[7,8,9]. Frontoethmoidalencephaloceles manifest as a clinically visible swelling over the bridge of the nose with significant hypertelorism and orbital deformity while the nasopharyngeal types remain occult and present with CSF rhinorrhea[4].Less common presentations include proptosis, meningitis, enlarging head sizeandneurofibromatosis[5]. The intracranial root of most frontoethmoidalencephaloceles lies at the foramen cecum, a small ostium located at the bottom of a small depression anterior to the crista galli and formed by the closure of the frontal and ethmoid bone.

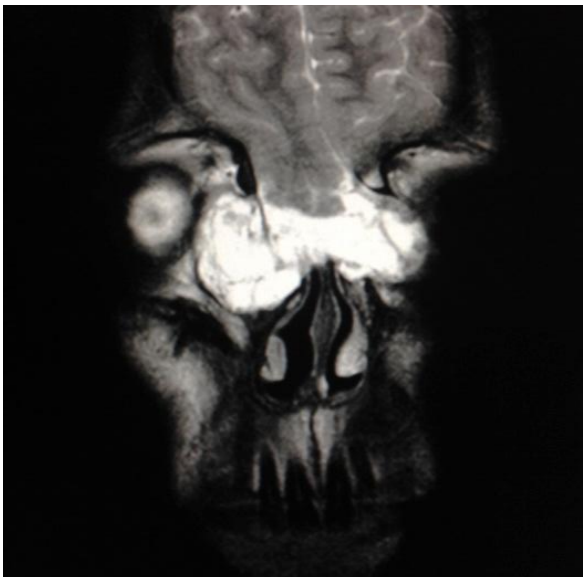


Fig 1: Coronal T2 W MRI Showing Defect in the anterior cranial fossa with herniation of meninges, CSF spaces and brain parenchyma into the nasal cavity and orbit reconstructed contrast enhanced CT scam abdomen shows bilateral diffuse enlargement of kidneys with heterogenous enhancement. No mediastinal or abdominopelvic lymphadenopathy was noted.

Although occipital encephaloceles may be associated with Chiari or Dandy-Walker malformations, Meckel Gruber Syndrome and callosal or migrational anomalies like hetertopias[10], frontoethmoidal lesions are not typically associated with these types of anomalies. Some studies report the associated findings on imaging were hydrocephalus (15%), corpus callosum agenesis (7%), arachnoid cyst (3%), porencephalic cyst (3%) and single ventricle (3%).MR imaging is the imaging modality of choice for defining the contents of an encephalocele for surgical planning. High-resolution CT may also be used to display the bone anatomy, but the intracranial connection and extent of cerebral tissue in the encephalocele arebest defined with MR imaging which determinesprognosis and surgical planning. Any associated intracranial anomalies and underlying brain gliosis are best seen with MRI . MRI is very useful in differentiating a frontoethmoidalencephalocele from a nasal glioma or nasal dermoid cyst. Frotoethmoidalencephaloceles are usually repaired with bilateral orbital advancement or hemi orbial advancement.

Postoperative CSF leak is the major complication reported in 10-15% cases[10,11,12]. However, by and large, CSF leak is transient and subsides with conservative management and repeated CSF drainage from lumbar theca[10,11,12].

The prognosis is variable dependent on the presence of associated anomalies and presence of microcephaly (carries a much poorer prognosis).

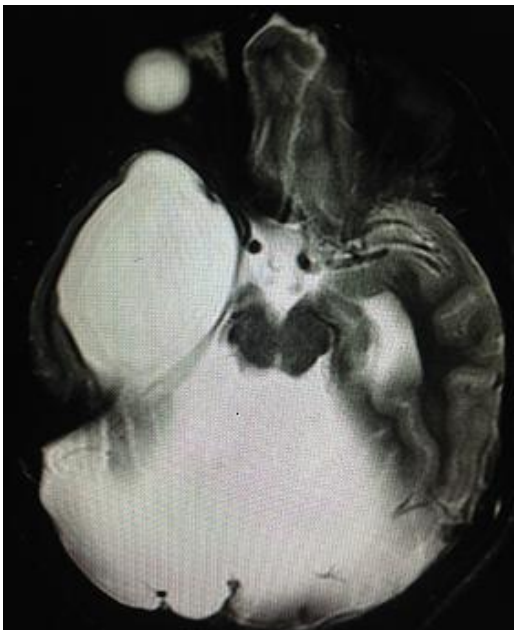


Fig 2: Axial T2 W MRI showing huge CSF intensity arachnoid cyst occupying the middle and posterior cranial fossa with significant mass effect on surrounding structures.

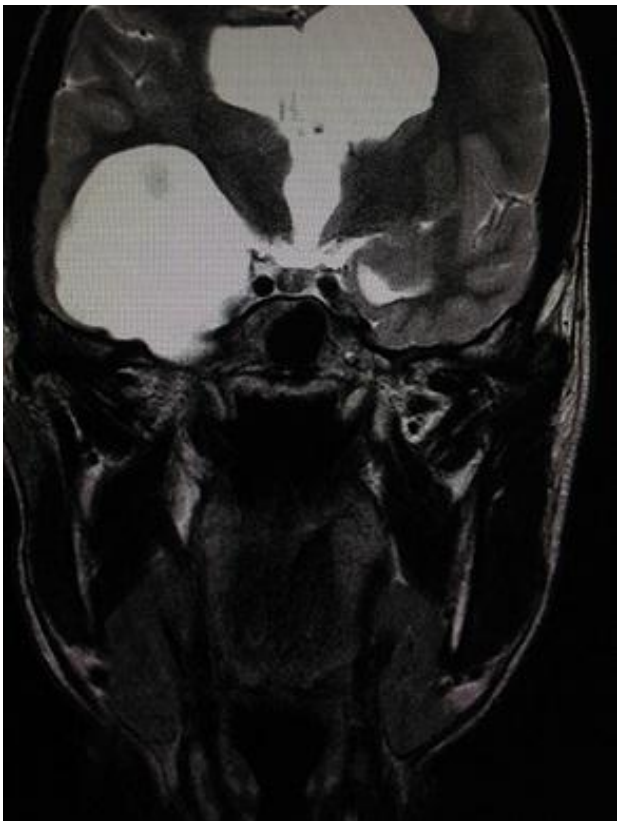


Fig 3: Coronal T2 W MRI revealing large arachnoid cyst in middle cranial fossa and mass effect with hydrocephalus.

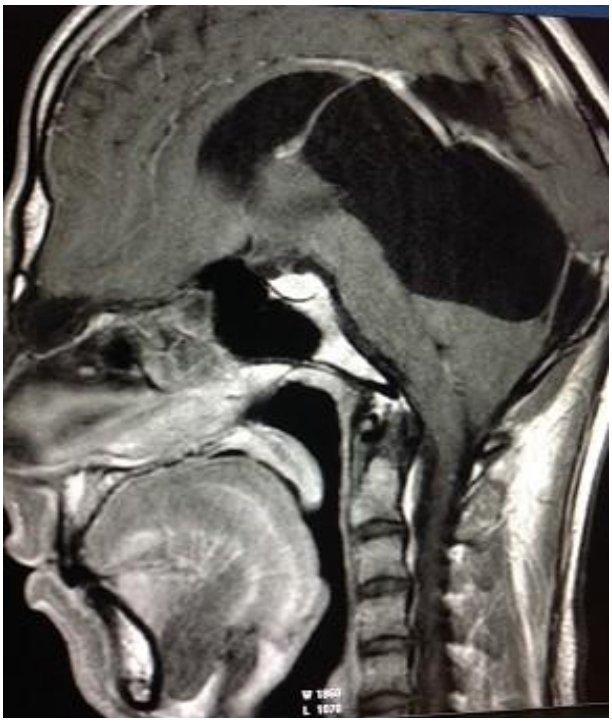
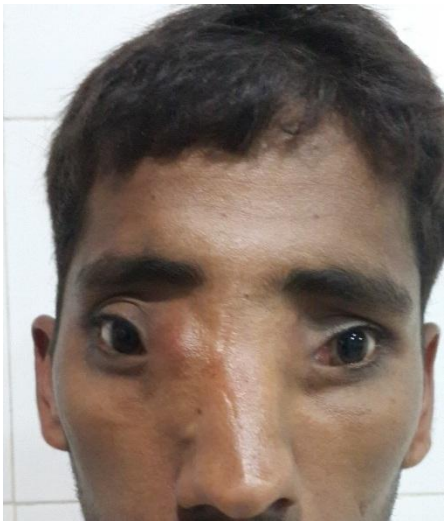


Fig 4: Sagittal T1 W MRI showing continuation of arachnoid cyst through the tentorium cerebelli into the posterior cranial fossa and mass effect on cerebellar hemispheres.



Case with encephlocoele

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