

## Rare case of a large congenital parietal meningoencephalocele associated with open lip schizencephaly

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**Section:** Paediatric radiology

**Area of Interest:** Neuroradiology brain

**Procedure:** Localisation

**Procedure:** Education

**Imaging Technique:** MR

**Special Focus:** Pathology Congenital Case Type: Clinical Cases

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**Patient:** 1 years, male

### Clinical History:

A one-year-old male child presented with a midline cranial swelling present since birth. The patient had had occasional convulsions for the past 6 months. He had been provisionally diagnosed with an encephalocele antenatally, and brain MRI was performed for preoperative evaluation.

### Imaging Findings:

Figure 1 (3D volume-rendered CT image) shows a midline scalp swelling.

MRI findings include a large parietal meningoencephalocele, with herniation of CSF and dysmorphic brain parenchyma through a midline skull vault defect measuring 5.2 x 4.3 cm. The encephalocele itself measures approximately 7.2 x 9.0 x 9.4 cm (AP x TR x CC). (Figures 2, 3, 4, 5 and 7.)

Further associated anomalies were also demonstrated, including partial agenesis of the posterior body and splenium of the corpus callosum (Fig. 2, 6 and 7), underdeveloped frontal horns of both lateral ventricles (Fig. 6), and a dilated third ventricle (Figures 2 and 7).

A unilateral parietal open lip schizencephaly with CSF-filled cleft was also demonstrated, with dysplastic thickened grey matter (Figures 7, 8 and 9) and polymicrogyria complex (Figure 7) adjacent to it.

There is involvement of the superior sagittal sinus within the meningoencephalocele. (Figure 4).

### Discussion:

Congenital parietal meningoencephalocele is a rare variant of meningoencephalocele, being a protrusion of meninges and brain parenchyma through a congenital midline skull vault defect. Meningoencephaloceles may be congenital or acquired, the latter being traumatic or postsurgical. Congenital meningoencephalocele occurs in 1-3 per 10, 000 live births, out of which parietal meningoencephalocele accounts for 5 to 10% of cases. It occurs due to

failure of complete closure of the neural tube during fetal life. Parietal meningoencephalocele arises from a skull defect between lambda and bregma. It is usually associated with other brain anomalies such as corpus callosum agenesis, Dandy-Walker malformation, Chiari II malformation and lobar holoprosencephaly [1, 2]. Here it is associated with schizencephaly, partial agenesis of the posterior body and splenium of the corpus callosum, with underdeveloped frontal horns of both lateral ventricles and third ventricle dilatation. A few published cases suggest an association with meningoencephalocele and schizencephaly [1]. There is a variant called atretic parietal encephalocele, which are small lesions under the scalp consisting of dysplastic brain tissue, fibrous tissue and dura [3].

Schizencephaly is a different entity, representing a cleft in cerebral parenchyma, lined by grey matter, and extending from ependyma to pia mater. It is a developmental disorder of supra-tentorial neuronal migration. Schizencephaly is a rare disorder that occurs in 1-2 per 1, 000, 000 births. There are two morphological types, called open lip when there is separation of the cleft which is filled with CSF, and closed lip when then cleft walls are opposed. Schizencephaly may be associated with other anomalies such as polymicrogyria, agenesis of the corpus callosum, grey matter heterotopia, and absence of the septum pellucidum. In our case it is associated with meningoencephalocele, polymicrogyria and partial agenesis of corpus callosum [4].

Both meningoencephalocele and schizencephaly may present with convulsions, motor impairment, hemiparesis and developmental delay [1-6].

Outcome depends upon the type of encephalocele and schizencephaly, whether there is involvement of superior sagittal sinus within the encephalocele, and on associated brain anomalies. Involvement of the superior sagittal sinus means neurosurgical repair is more difficult. Parietal meningoencephalocele is usually associated with structural brain anomalies and therefore has a poorer prognosis.

Management is usually symptomatic treatment for seizures, and surgery and VP shunt for hydrocephalus. Further management may include physiotherapy and occupational therapy [1-6].

**Differential Diagnosis List:** Congenital parietal meningoencephalocele with unilateral open lip schizencephaly, Hydrocephalus, Porencephaly

**Final Diagnosis:** Congenital parietal meningoencephalocele with unilateral open lip schizencephaly

## References:

- Mishra SS, et al. Large vertex meningoencephalocele with schizencephaly: An interesting case with neurosurgical challenge. J Pediatr Neurosci. 2014 May-Aug, Volume 9, Issue 2, Page 136-138 (PMID: [25250068](#))
- Gao Z, Massimi L, Rogerio S, et al. Vertex cephaloceles: A review. Childs Nerv Syst 2014; 30: 65–72. (PMID: [23989428](#))
- Siong Lung Wong, Huong Ling Law, and Suzet Tan Atretic Cephalocele — An Uncommon Cause of Cystic Scalp Mass. Malays J Med Sci. 2010 Jul-Sep; 17(3): 61–63. (PMID: [22135551](#))
- N. Hayashi, Y. Tsutsumi, A.J. Barkovich Morphological features and associated anomalies of schizencephaly in the clinical population: detailed analysis of MR images. Neuroradiology 2002, May, Volume 44, Issue 5, pp 418–427 (PMID: [12012128](#))
- Fanny E. Moro ?n, Michael C. Morriss, Jeremy J. Jones, Jill V. Hunter, Lumps and Bumps on the Head in Children: Use of CT and MR Imaging in Solving the Clinical Diagnostic Dilemma. Radiographics 2004; 24: 1655-1674 (PMID: [15537975](#))
- Nayak A, Sharma S, Vadher RK, Dixit S, Batra RS. Congenital interparietal encephalocele: a case report. J Clin Diagn Resv. 9(4); 2015, April, PD09–PD10, PMC4437110 (PMID: [26023594](#))

**Figure 1**

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**Description:** 3D volume rendered CT image demonstrates a large midline scalp swelling. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 2**

a



**Description:** Axial T2W image of brain shows underdeveloped frontal horns of bilateral lateral ventricles. Non-visualisation of splenium of corpus callosum suggests partial agenesis of corpus callosum. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 3**

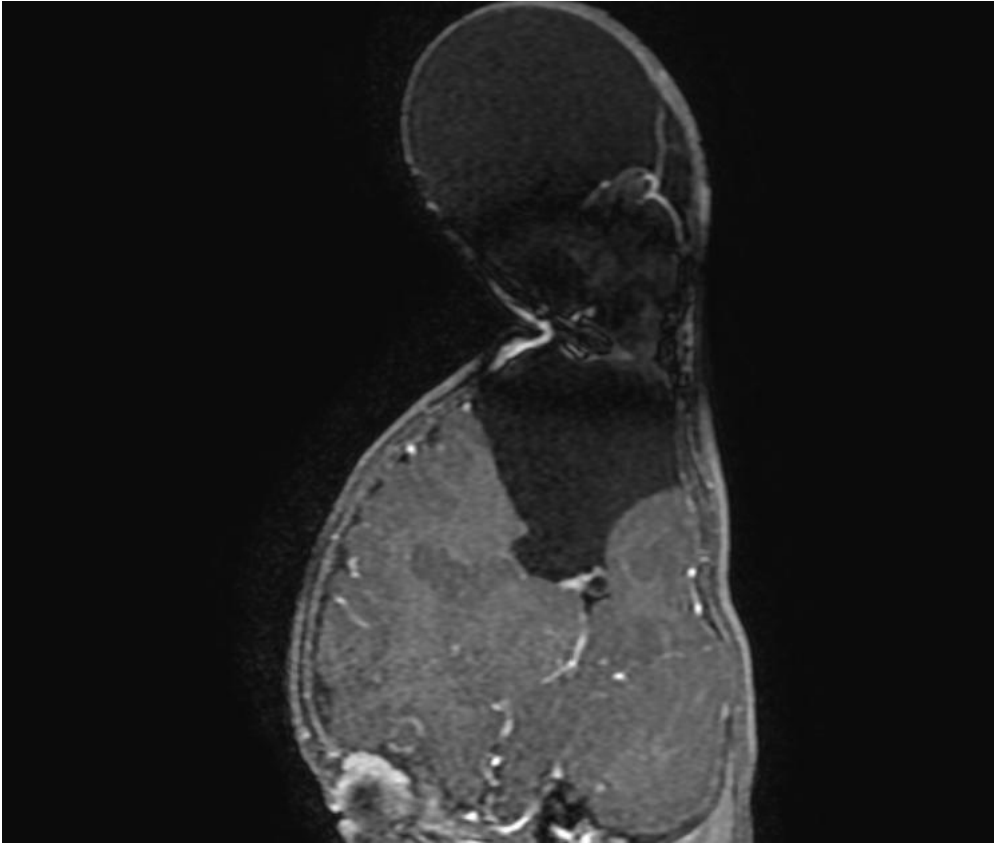
a



**Description:** Midline sagittal T1W post-contrast image of brain shows polymicrogyria complex adjacent to open lip schizencephaly. Dilated third ventricle and partial agenesis of the posterior body & splenium of corpus callosum is noted. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 4**

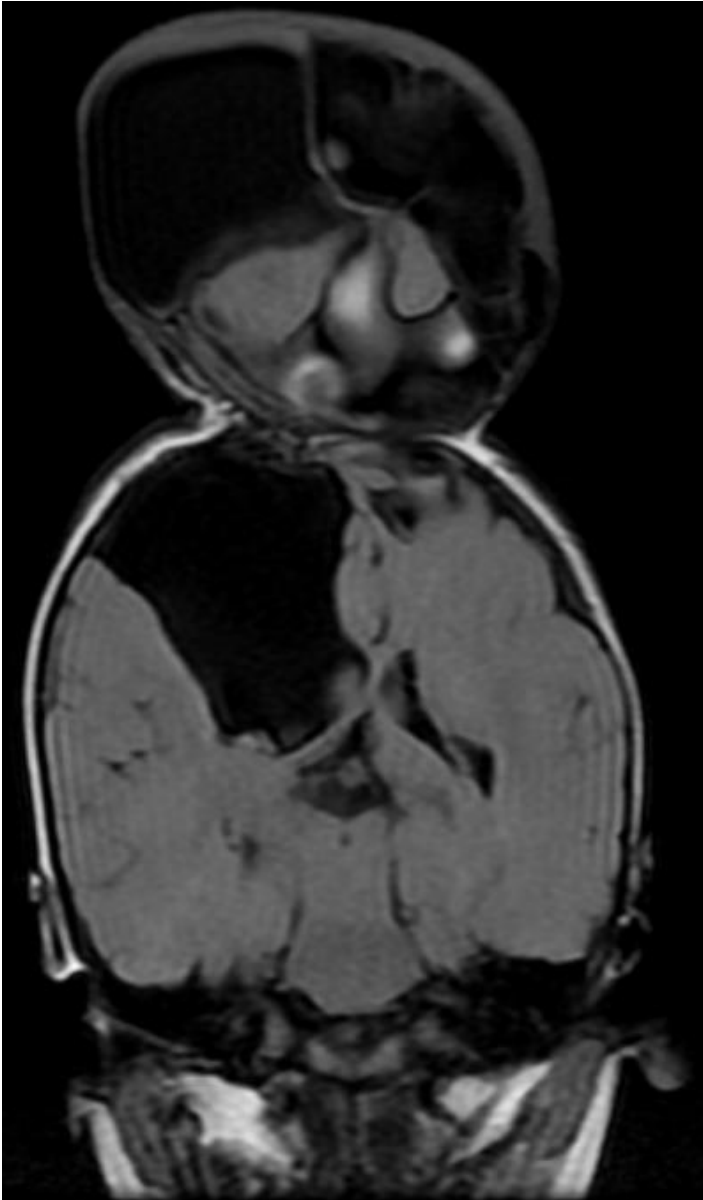
**a**



**Description:** Sagittal T1W post-contrast image shows a parietal open lip schizencephaly with dysplastic thickened grey matter adjacent to it. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 5**

a



**Description:** Coronal T2W FLAIR image shows large parietal meningoencephalocele as evidenced by herniation of CSF and dysmorphic brain parenchyma through midline skull vault defect. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 6**

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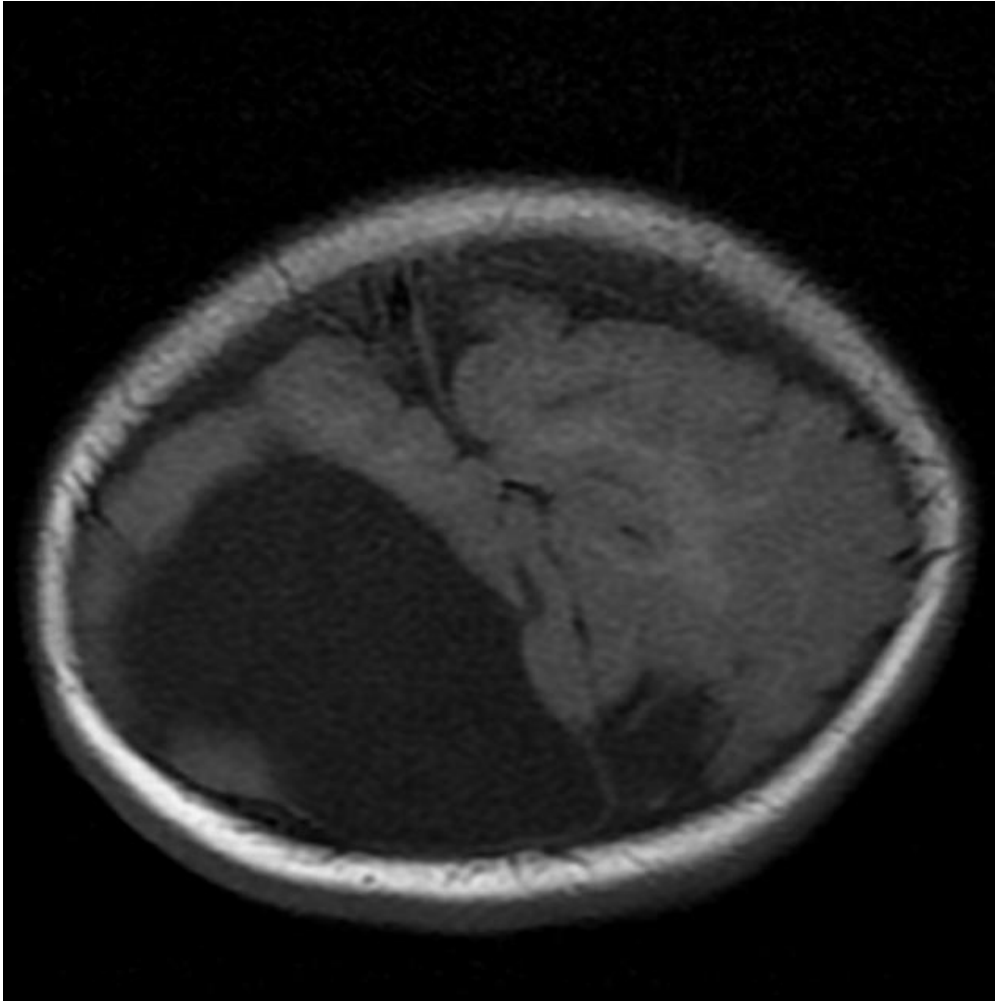


**Description:** Sagittal T1 Inversion Recovery Image shows partial agenesis of the posterior body & splenium of corpus callosum, meningoencephalocele and dilated third ventricle. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.



**Figure 7**

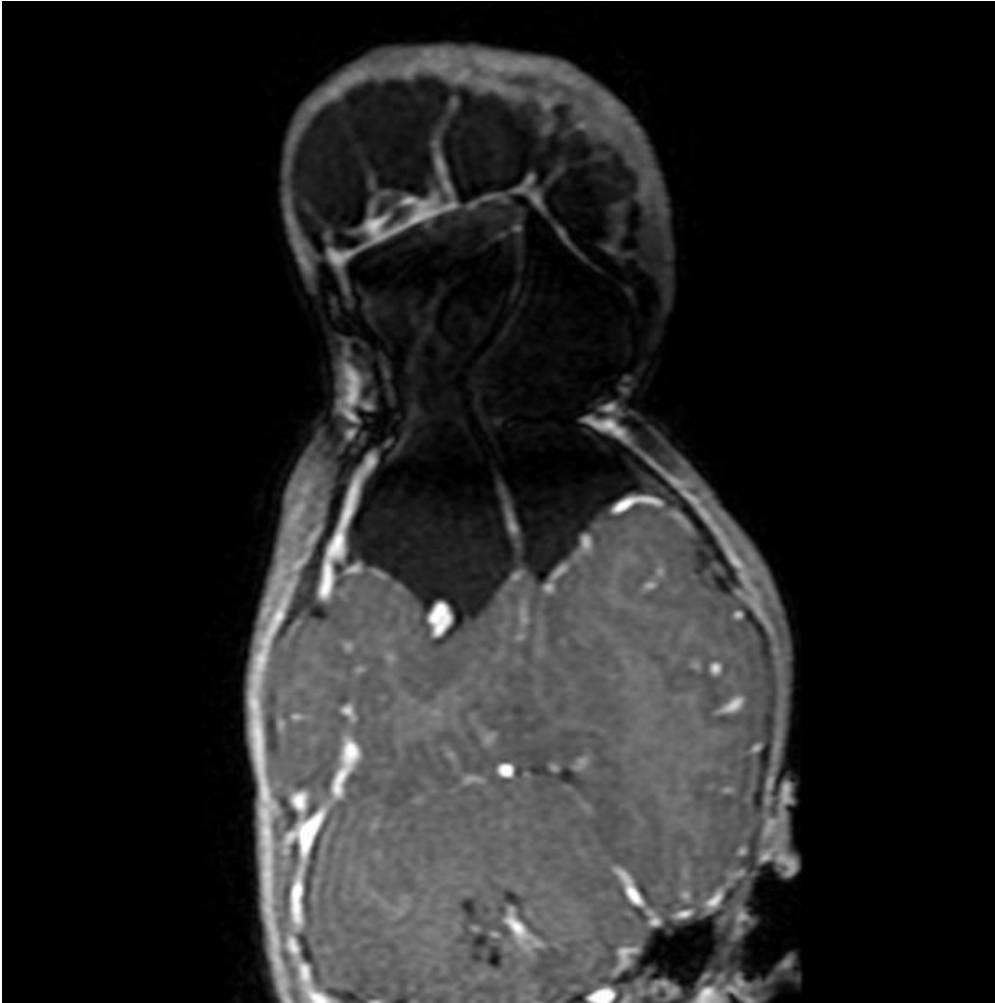
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**Description:** Axial T1W image shows unilateral parietal open lip schizencephaly filled with CSF. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 8**

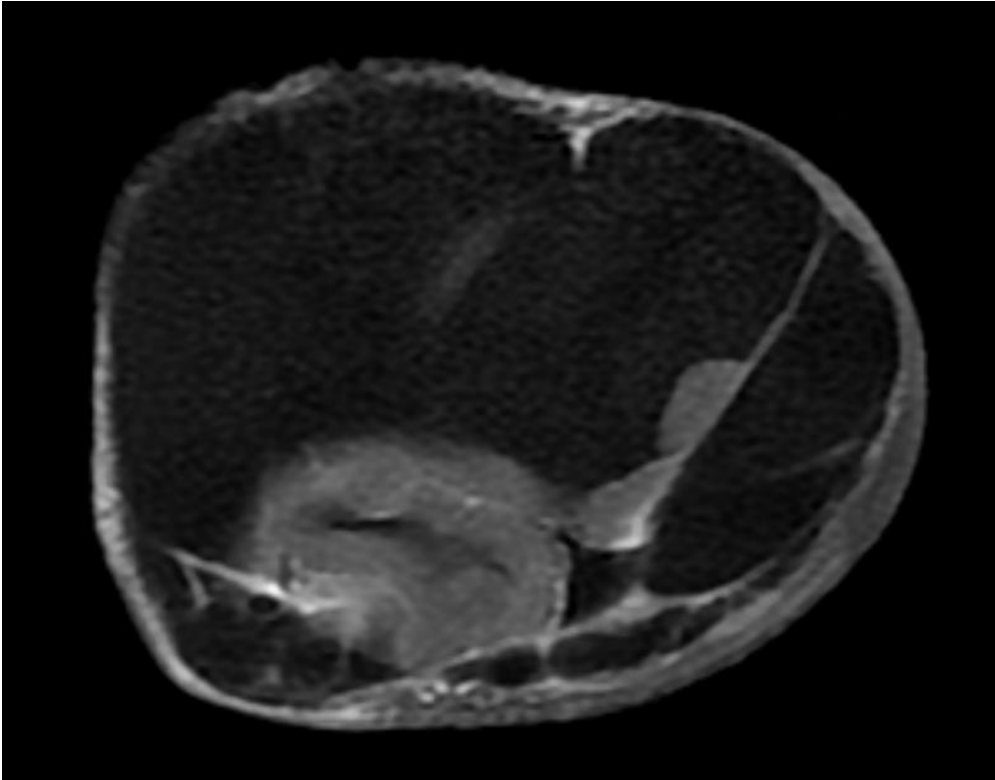
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**Description:** Coronal T1W post-contrast image shows involvement of superior sagittal sinus and meningoencephalocele. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.

**Figure 9**

**a**



**Description:** Axial T1W post-contrast image at the level of parietal meningoencephalocele. **Origin:** Department of Radiology, P.D.U. Medical college, Rajkot.