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CASE REPORT

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Unusual pattern of leptomeningeal cyst with herniation of porencephalic cyst

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Abstract

Background Growing skull fracture or leptomeningeal cyst is an uncommon occurrence after severe head trauma in childhood. It is mostly observed in infants and children less than 3 years of age. Another uncommon complication of head trauma is development of porencephalic cyst.

Case presentation We present an unusual case of post-traumatic type III leptomeningeal cyst in a 9-month-old infant with history of head trauma 3 months ago. CT and MR imaging revealed widening of bony defect compared to previous imaging, and herniation of porencephalic cyst through the defect, leading to formation of a large cystic swelling in scalp. The 3 month delay in evaluation of the scalp swelling was due to lack of patient education after trauma, and no subsequent follow-up.

Conclusions Knowledge about etiopathogenesis and risk factors of leptomeningeal cyst development after head trauma ensures that close follow-up is done in such cases for early detection and management of growing skull fracture.

Keywords Growing skull fracture, Head trauma, Leptomeningeal cyst, Trauma

Background

Development of leptomeningeal cyst after trauma in children is an uncommon occurrence. It involves gradual widening of calvarial defect and herniation of intracranial contents through the defect. [1]. It is an important differential for an infant or child presenting with scalp swelling. History of recent head trauma and clinical examination revealing pulsatile scalp swelling are essential components of this lesion. Imaging modalities helpful in diagnosis of leptomeningeal cyst include X-Ray, CT and MRI. While CT and X-Ray help in delineating the cortical defect in skull, MRI reveals the dural defect and herniated intracranial contents [2]. Early recognition of leptomeningeal cyst by identification of at-risk individuals by frequent follow-ups after trauma is important for good prognosis. In this case report, we present an usual pattern of leptomeningeal cyst with herniation of porencephalic cyst.

Case presentation

A 9-month-old infant was brought to our institute with complaint of pulsatile swelling in occipital region, progressively increasing in size. The swelling started after the child fell from height 3 months back. Initially, the swelling was small (3 cm), and the size gradually increased over one month to current size (approx. 10 cm). On physical examination, the swelling was cystic in consistency.

Non-contrast computed tomography of head was performed after the fall revealed a linear partially displaced fracture with diastasis of 4 mm in occipital bone involving both outer and inner tables along with haemorrhagic contusion in right parietooccipital region (Fig. 1). The child was managed conservatively. The patient did not report for regular follow-up and presented later with a large occipital swelling.

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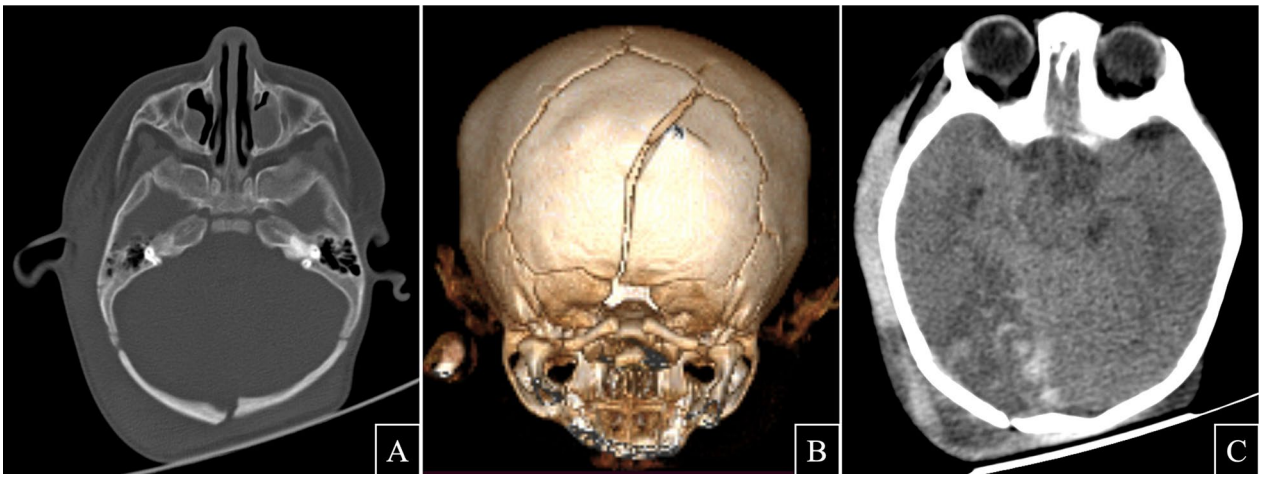

Fig. 1 A 9-month-old infant with head injury after fall—Non-Contrast CT scan **a** axial image in bone window, **b** VRT image reveals partially displaced fracture in the occipital bone extending up to foramen magnum and right parietal bone. **c** Axial image in brain window shows haemorrhagic contusion in right parietooccipital region along with heterogeneous swelling in scalp in occipital region

Present CT head revealed widening of calvarial defect (3.3 cm) with everted edges of inner table along with hypodense fluid attenuation area in right parietooccipital region which was communicating extracranially with large cystic swelling in scalp. MRI of the brain was performed for further characterisation. It revealed a large porencephalic cyst (communicating with occipital horn of right lateral ventricle) in right parietooccipital lobe herniating extracranially via defect in occipital bone and underlying dura (Fig. 2). Chronic haemorrhage with encephalomalacic and gliotic changes were also seen in right parietal lobe.

Thus, a diagnosis of type III leptomeningeal cyst with herniation of porencephalic cyst was made. The patient was managed surgically, with craniectomy and watertight duraplasty.

Discussion

Growing skull fracture or leptomeningeal cyst is an uncommon occurrence after severe head trauma in childhood. It is mostly observed in infants and children less than 3 years of age. Cases in children more than eight years or adults are rare [2]. Both skull fracture and dural laceration are essential elements for pathogenesis of growing skull fracture [3]. A head injury causing haemorrhagic contusion is considered sufficient to cause dural laceration and thus, can serve as a marker for future risk of leptomeningeal cyst. Similarly, fracture diastasis of > 4 mm is an additional risk factor for development of leptomeningeal cyst. Factors responsible for development of growing skull fracture in children include adherence of dura mater to skull in children and rapid growth of skull in first two years of life [4].

In older individuals, linear fracture in thin area of skull base like orbital roof, ethmoid plate, frontal sinus along with dural laceration can lead to leptomeningeal cyst formation [5].

Clinically, growing skull fracture presents with progressive, pulsative scalp mass. Neurological complaints include seizures, hemiparesis, psychomotor retardation [6]. In our case, although the infant was brought to the hospital after 3 months, no neurological symptom had reported.

Three type of leptomeningeal cysts have been described based on imaging findings. Type I involves herniation of subarachnoid space fluid, while type II involves herniation of gliotic brain parenchyma. Type III is the unusual pattern in which porencephalic cyst communicating with ventricular system is herniated through the bony calvarium. [7]

CT carried out before the development of leptomeningeal cyst delineates the fracture pattern and associated haemorrhagic contusions. Widening of fracture line, everted edges of fracture and herniation of intracranial contents are the signs of leptomeningeal cyst formation on CT imaging. MRI provides better evaluation of dural tear and herniated intracranial contents [1, 3]. In our case, widening of fracture line and everted fracture edges were evident on CT. However, the diagnosis of herniated porencephalic cyst communicating with ventricular system was diagnosed on MRI based on the flow void between cyst and lateral ventricle (Fig. 2a).

Liu et al. [8] described the stages of growing skull fracture for treatment planning. Stage I begins after the initial trauma to just before fracture expansion. Stage II follows initial fracture enlargement, with entrapped

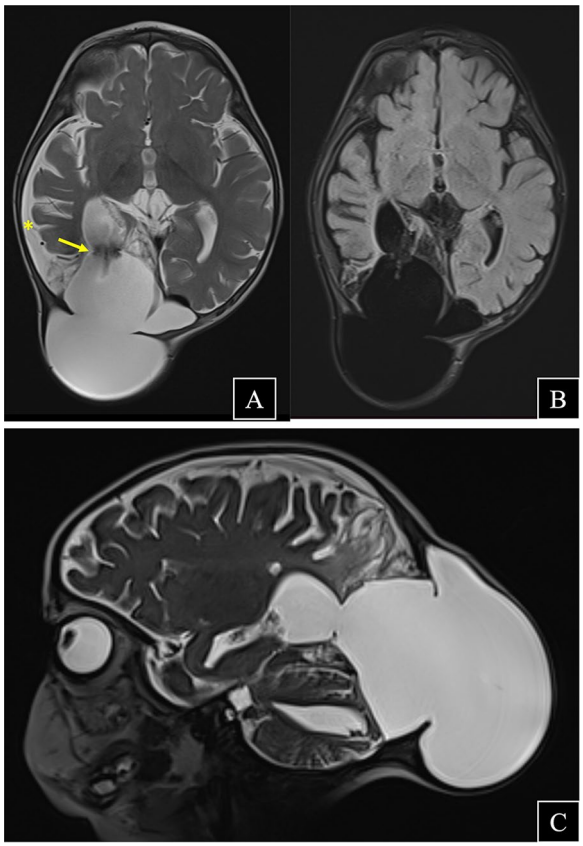

Fig. 2 MRI of the brain carried out 3 months after head trauma due to complaint of occipital swelling **a** Axial T2 weighted TSE, **b** FLAIR images show porencephalic cyst in right parietooccipital region with flow void between cyst and occipital horn of right lateral ventricle. (arrow) Extracranial communication of porencephalic cyst with scalp swelling noted via wide defect in the occipital bone with everted edges. Chronic subdural hematoma is seen in right temporal region (*). **c** Sagittal T2 SPACE image shows porencephalic cyst communicating with ventricle and extracranial cyst

tissue in skull fracture. Stage III (late stage growing skull fracture) shows significant enlargement of calvarial defect with extracranial herniation of brain and CSF.

Treatment strategies for growing skull fracture include VP shunt for management of hydrocephalus, craniotomy or cranioplasty along with duraplasty. Dural repair forms the cornerstone of treatment, and patient should be monitored for raised intracranial pressure after watertight duraplasty [1, 8].

Early recognition and management of growing skull fracture reduce future complications and lead to more favourable results [4]. Knowledge about growing skull fractures and its risk factors is essential for identification of possible future complications. Clinical follow-up is required over next several weeks for such cases and should focus on development of neurological deficit and monitoring the size of hematoma/ scalp swelling.

Conclusions

Growing skull fracture should be considered as potential complication of severe head trauma in childhood. Close follow-up and identification of early symptoms and signs enables early diagnosis and management of growing skull fracture.

Abbreviations

CT	Computed tomography
MRI	Magnetic resonance imaging
CSF	Cerebrospinal fluid

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Author contributions

Both authors performed the CT, MR acquisition. KV performed the post processing of MRI images. SA performed the literature search and prepared the manuscript. Both authors prepared the figures and figure legends. Both authors reviewed and edited the manuscript. Both authors read and approved the final manuscript.

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Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Waived off by Institute Ethics Committee, Dr RML Hospital, New Delhi, in view of case report. Informed written consent was taken from the participant.

Consent for publication

Informed written consent was taken from the participant.

Competing interests

The authors declare that they have no competing interests.

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