Case Report

A rare neglected case of large leptomeningeal cyst: A case report

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ABSTRACT

Leptomeningeal cysts are enlarging skull fractures that occur near post-traumatic encephalomalacia. The term cyst is usually a misnomer as it is not only a cyst but also an extension of encephalomalacia. They commonly occur after severe head trauma during the first 3 years of life (especially infancy) and almost never after 8 years of life. Herewith, we present a case of a 7-year-old female patient who presented to our hospital with left-sided weakness since 2 years and painless swelling over the scalp since 3 years giving past history of head trauma. Plain radiography, computed tomography, and magnetic resonance imaging were performed which showed classical findings of leptomeningeal cyst. We will be discussing about the causes and radiological findings which help in diagnosis as well as treatment planning.

Key words: *Computed tomography, Encephalomalacia, Leptomeningeal cyst, Magnetic resonance imaging*

Skull fractures are either linear or depressed. If a skull fracture involves meninges, there occurs movement of cerebrospinal fluid (CSF) from the subarachnoid space. It collects and leads to localized cyst formation due to pressure erosion "leptomeningeal cyst" which leads to bone atrophy and increasing defect. Linear fractures are typically <3 mm, wider in midportion, and narrower at ends. They commonly occur after severe head trauma during the first 3 years of life (especially infancy) and almost never after 8 years of life. We are presenting a case which was neglected for many years without any medical help.

CASE REPORT

A 7-year-old female patient presented to our hospital with left-sided weakness for 2 years and painless swelling over the scalp since 3 years. There was a history of head trauma 6 years back when she was banged against the wall by her father at the age of 1 year. However, she was not brought to any hospital, and no investigation or management was done at that time. There was no history of unconsciousness, convulsions, chronic headache, and recurrent vomiting or fever in the past.

On examination, she was conscious, well oriented and her vitals were within normal limits. The local examination revealed soft swelling of the scalp on the left side, which was soft, and non-tender with normal skin, and no local rise in temperature. No other significant clinical findings were present. Left-sided hemiparesis was present, and rest of the systemic examination was normal. Fundus examination was normal. A provisional diagnosis of cystic encephalomalacia was given, and the patient was referred to the our department for radiological investigations.

Plain radiograph, lateral view of the skull revealed a lytic lesion with thick irregular sclerotic margins in the left temporo-parieto-occipital region (Fig. 1a). Computed tomography (CT) by 128 slice Siemens perspective equipment showed the cystic lesion passing through erosion of temporal, parietal, and occipital bones and extending within the calvaria and extracalvarially and causing calvarial widening measuring 1.5 cm with beveled edges. The defect in the calvaria measured around 4.5 cm. There was no herniation of brain parenchyma seen at this place. The cyst was communicating with the ipsilateral dilated occipital horn (20 mm) of the lateral ventricle and causing compression over the ipsilateral temporo-occipital lobe (Figs. 1 and 2). There were prominent sulci, cisterns, and ventricles. The temporal horn of lateral ventricle was dilated 12 mm; however, other ventricles were normal in size.

Imaging by 1.5 Tesla magnetic resonance imaging (MRI), Siemens Avento revealed: A well-defined T1 and FLAIR hypointense and T2 hyperintense cystic CSF signal intensity/density lesion measuring 6.5 × 6 × 4.5 cm (AP × TR × CC) and volume of 100 cc in the left temporo-parieto-occipital region (Fig. 3). With the above findings, diagnosis of leptomeningeal cyst was made. Parents were counseled about the surgical options, but they refused for the surgery.

DISCUSSION

Leptomeningeal cyst or growing skull fracture was first described by Howship in 1816. Leptomeningeal cyst is an enlarging skull fracture that occurs near post-traumatic encephalomalacia. The term cyst is usually a misnomer as it is not only a cyst but also

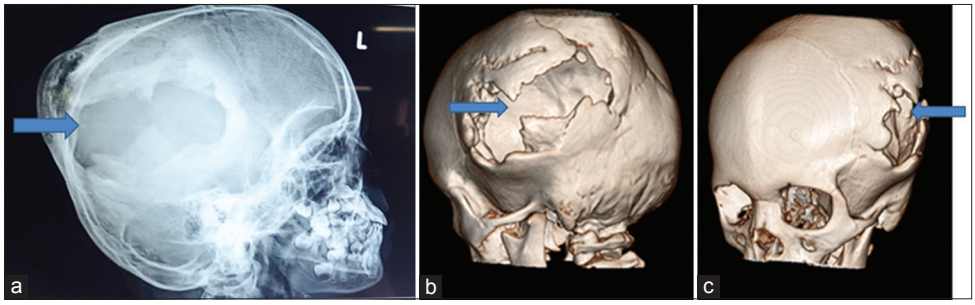

Figure 1: (a-c) Skull - lateral view and three-dimensional reconstruction computed tomography images showing a lytic lesion (arrow) with thin irregular sclerotic margins with beveled edges in the left temporo-parieto-occipital region

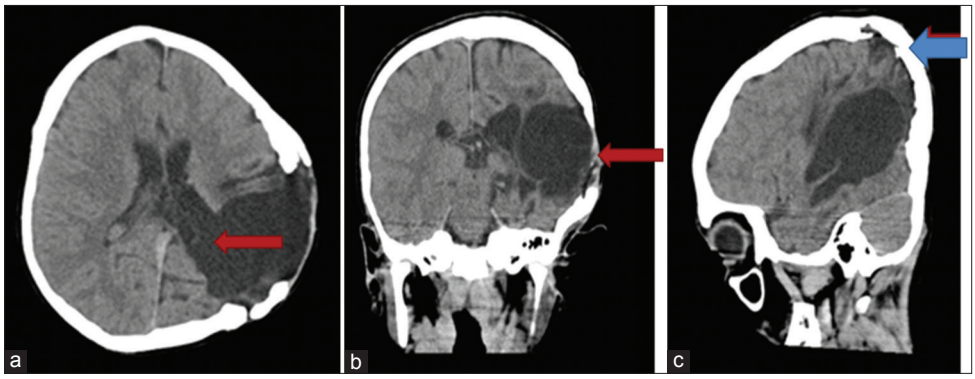

Figure 2: (a-c) Plain computed tomography axial (a) and coronal images (b) showing hypodense lesion (arrows). Lesion is communicating with the lateral ventricle suggestive of porencephalic cyst. (c) Sagittal image shows hypodense lesion causing pressure erosion over the (arrows)

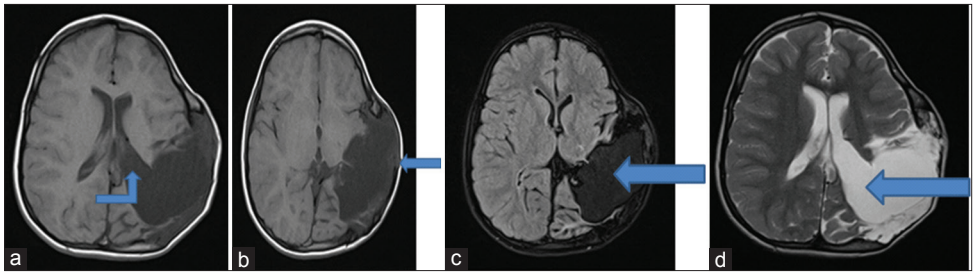

Figure 3: (a-d) T1-weighted axial magnetic resources images (MRI) showing a hypointense lesion protruding (arrow) and porencephalic cyst. (d) T2-weighted axial MRI showing hyperintensity

an extension of encephalomalacia. Hence, it is usually seen a few months post trauma [1-3]. The incidence rate is 0.05-0.1% of the skull fractures in childhood. Growing skull fracture usually occurs after severe head trauma during the first 3 years (especially infancy) and almost never after 8 years of life, as during this age, the brain volume increases rapidly which is in part responsible for its development. Most common site is parietal region [3,4].

Patient presents with progressive, often pulsatile, scalp mass that appears usually after head trauma sustained after infancy. Associated symptoms include asymptomatic enlarging palpable mass, seizures, hemiparesis, psychomotor retardation, and focal neurological deficit. There are multiple factors which are responsible for its development such as (a) CSF pulsations, (b) brain trauma with dura mater laceration, and (c) craniotomy without proper dural laceration repair leads to fracture in the thin skull to enlarge and it keep the tear open, which prevents healing of both the dura and the fracture [4]. Latter fracture margins become progressively widened, bevelled, and sclerotic.

A growing fracture with leptomeningeal cyst formation is more commonly seen in falls, child abuse, physical assaults, and road traffic accidents [1]. The brain extrusion may be present shortly after diastatic linear fracture in neonates and young infants resulting in focal dilatation of the lateral ventricle near the growing fracture. This focal dilatation is reversible and may normalize after surgical repair. For growing fractures to develop, a dural laceration is a must with a fracture line. Cranial defects never increase if the underlying dura is intact. Another risk factor is the severity of underlying trauma [4].

A linear fracture associated with a hemorrhagic contusion of adjacent brain suggests a trauma significant enough to cause dural laceration. The brain at the growing fracture site shows a cerebromeningeal cicatrix formation. Cystic changes at the growing fracture site are because of cystic encephalomalacia. These skull fractures after reaching their maximum extent cease to grow and remain stable throughout childhood [2].

A fracture with a diastasis of >4 mm may be considered at risk of developing a growing skull fracture [4]. In older individuals, skull fracture healing time in adults is usually 6-8 years. Growing

fractures can even be seen in older individuals usually in linear fractures in thin areas of skull base associated with dural laceration, for example, orbital roof, ethmoid plate, and frontal sinus [4]. Pathologically, tissue interposition because of fracture prevents osteoblasts from migrating to the fracture site, inhibiting healing, and the resorption of the adjacent bone by the continuous pressure from tissue herniation through the bone gap adds to the progression of the fracture line.

CT is the imaging modality of choice as it shows a clear depiction of bony as well as cerebral changes. It consists of a lytic calvarial lesion with scalloped edges, in which encephalomalacia invaginates. Other features may also be present such as extracranial brain herniation, hydrocephalus, unilateral ventricular dilatation, and porencephalic cyst [3,5]. Cranial defects never increase if the underlying dura is intact. Brain extrusion may be present resulting in focal dilatation of the lateral ventricle near the growing fracture.

Prompt diagnosis is essential as early corrective surgery should be performed for growing skull fractures so as to prevent secondary brain damage and neurological deficits which, especially, occur in late stages and lead to progressive disruption in the quality of life of patients. Surgical treatment involving excision of meningocele and repair of bone and dural defects is associated with a good outcome. Dural defect needs to be fully repaired. In our case, it was neglected for a very long time, so surgical options with damage to brain development become a tough call.

CONCLUSION

Leptomeningeal cysts are enlarging skull fracture that occurs near post-traumatic encephalomalacia and usually occurs after severe head trauma during the first 3 years of life. In infants and children, linear skull fractures should be monitored until definite skull bone consolidation so as to prevent progression and they should not be left neglected.

REFERENCES

1. Ersahin Y, Gülmen V, Palali I, Mutluer S. Growing skull fractures (craniocerebral erosion). Neurosurg Rev. 2000;23:139-44.
2. Hamamcioglu MK, Hicdonmez T, Kilincer C, Cobanoglu S. Large intradiploic growing skull fracture of the posterior fossa. Pediatr Radiol. 2006;36(1):68-70.
3. Kim H, Jo KW. Treatment of a traumatic leptomeningeal cyst in an adult with fibrinogen-based collagen. J Korean Neurosurg Soc. 2013;53(5):300-2.
4. Khandelwal S, Sharma GL, Gopal S, Sakhi P. Growing skull fractures/ leptomeningeal cyst. Indian J Radiol. 2000;23:139-44.
5. Sutton D. Textbook of Radiology and Imaging. 7th ed., Vol. 2. New York: Churchill Livingstone; 2003. p. 1641-2.

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