

Case Report

Large Intradiploic Growing Skull Fracture of the Posterior Fossa with Syringomyelia

Abstract

Growing skull fracture (GSF), also known as leptomeningeal cyst, is a rare but significant complication of pediatric head injury. It is mainly seen in infancy and childhood. GSFs of the posterior fossa are uncommon, and intradiploic location in the posterior fossa is extremely rare. Only a few cases of pediatric GSF of the posterior fossa and intradiploic location have been reported in the literature. We report a case of a 15-year-old boy who had large intradiploic GSF of the posterior fossa, associated with cervical syringomyelia. The lesion developed progressively over a period of 5 years following a documented occipital linear fracture. This case of a GSF developing from a known occipital linear fracture demonstrates that a GSF may reach a considerable size, and although uncommon, intradiploic development and occipital localization of a GSF are possible. Syringomyelia associated with posterior fossa GSF is very unusual which makes this case even more unique.

Keywords: Intradiploic, leptomeningeal cyst, posterior fossa growing skull fracture, syringomyelia

Introduction

Growing skull fracture (GSF) is a progressively enlarging diastatic fracture associated with an underlying dural laceration. It is a rare complication of head injury. The incidence of GSF is reportedly in 0.05%–1.6% of pediatric skull fractures.^[1] They mostly occur in the first decade of life although occurrence in later decades has been reported.^[2-4] The posterior fossa is an uncommon location for the development of GSF, as it is intradiploic in nature. Only a few cases of pediatric GSF of the posterior fossa and intradiploic location have been reported in the literature.^[2,3,5-8]

We report a 15-year-old boy with a large posterior fossa intradiploic GSF associated with syringomyelia that developed progressively over 5 years after a documented linear occipital fracture.

Case Report

A 15-year-old boy presented with a complaint of progressively increasing swelling over the back of his head for 5 years. He also had dull aching occipital headache with giddiness of 2-week duration. His medical history revealed that 5 years earlier, he had sustained a linear occipital fracture due to a fall from a height.

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Birth history was normal. Neurological examination was within normal limits. The palpable occipital mass appeared shortly after the trauma, progressively enlarged over time, and reached the considerable size of 12 cm × 7 cm × 4 cm [Figure 1].

Computed tomography scan of the head [Figure 2] showed a 12 cm × 7 cm × 4 cm large-sized cerebrospinal fluid (CSF)-attenuated intradiploic lesion in the occipital bone. There was a severe thinning of the inner and outer table of the occipital bone. Bony defect was noted in the inner table in midline through which this lesion was communicating with fourth ventricle. Magnetic resonance imaging (MRI) of the head confirmed that the intradiploic lesion was filled with CSF and it was communicating with fourth ventricle [Figure 3a]. Screening of the cervical spine showed syringomyelia extending from craniovertebral junction (CVJ) to C3 vertebral body level [Figure 3b].

At surgery, the outer and inner table of the occipital bone was extremely thinned out along with evidence of CSF within the intradiploic space [Figure 4a and b]. Small linear defect of 1 cm × 5 mm was visualized in the inner table along with underlying dural defect through which

How to cite this article: Naik H, Velho V, Bhide A, Guha A. Large intradiploic growing skull fracture of the posterior fossa with syringomyelia. Asian J Neurosurg 2019;14:227-30.

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Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_275_17

Quick Response Code:





Figure 1: Clinical photograph of swelling

CSF was coming into the intradiploic space [Figure 4c]. Watertight repair of the dural defect was done using pericranium and then reinforced with fibrin glue [Figure 5a]. Defect of the outer table was repaired using Osteomesh [Figure 5b]. The patient recovered well without any fresh neurological deficit. MRI cervical spine done at 6-month follow-up showed a reduction in the size of syrinx [Figure 6a and b].

Discussion

GSF most commonly occurs in the parietal bone. It rarely occurs in the temporal or occipital bones probably because of the overlying muscles in these areas that prevent its development.^[2] Only a few previously reported pediatric cases of posterior fossa GSF were intradiploic in the location.^[2,3,5-8] Palaoglu *et al.*^[3] reported that leptomenigeal cysts involving the occipital bone represent a different radiological entity showing an intraosseous location. Posttraumatic intradiploic leptomenigeal cysts are extremely rare complications of calvarial fractures occurring in pediatric patients. These cysts are characterized by fracture of the inner table and laceration/tear of the dura mater with accumulation of CSF in a sac with a covering lined by arachnoid membrane and situated within the diploic space. Almost all patients have a history of trauma with the time interval between trauma and diagnosis of posttraumatic intradiploic leptomenigeal cyst being extremely variable. Clinical presentation may include a slow-growing swelling associated with headache, giddiness, and occasionally ataxia.^[9]

The most widely agreed-upon hypothesis proposes the herniation of leptomeninges into the intradiploic space through the traumatic rent in the dura mater and inner table. The ball and valve effect due to the child's growing brain and the continuous CSF pulsations act as driving expansile forces facilitating the growth of the intradiploic cyst over the years with resultant thinning of the outer

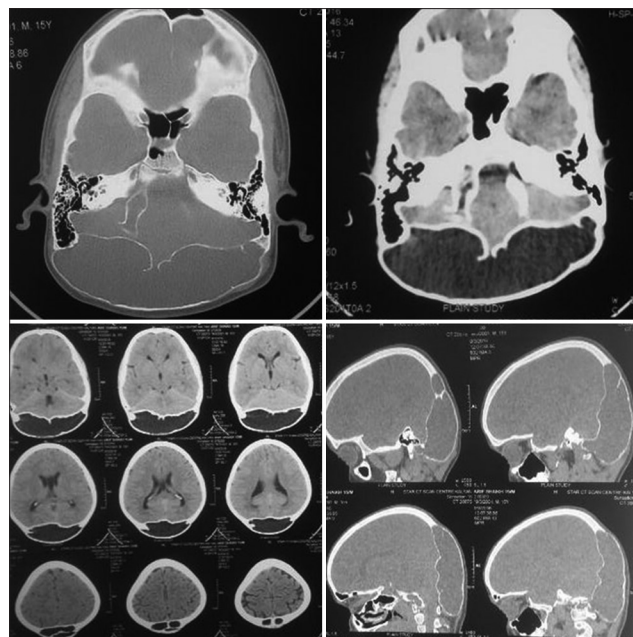


Figure 2: Axial and sagittal computed tomography images show the thin and bulging occipital external table resulting from the large, intradiploic cerebrospinal fluid density cyst. There was a severe thinning of the inner and outer table of the occipital bone. Bony defect was noted in the inner table in midline through which this lesion was communicating with fourth ventricle

table. The thickness of the occipital bone and the thick muscle cover buttressing its outer table explain the predilection of this entity to normally prevent occurring in the occipital region. Other contributory factors in this regard may include the more capacious diploic space as well as cartilaginous origin of the occipital bone compared to the membranous origin of the rest of the calvaria. Communicating type of hydrocephalus is often seen to occur along with posterior fossa GSF and may be attributed to the intraventricular hemorrhage at the time of initial trauma.

The common features of intradiploic GSFs are an intact outer table, a CSF-filled cyst, and a defect in the inner table and dura. The cysts are generally lined with arachnoid membrane.^[6] It has been argued that because of the actual thickness of the occipital bone, a fracture confined to the inner table and the associated dural laceration might result in leptomenigeal cyst formation.^[2,3,6] In our patient, however, the occipital linear fracture involved both inner and outer tables and was associated with syringomyelia, and thus the presented case differs from other cases reported in the literature.^[2,3,5-8]

Surgical repair of the dural and bony defect is the mainstay of treatment. It involves watertight duraplasty followed by cranioplasty using autologous split calvarial graft or Osteomesh. The technique of cranioplasty using Osteomesh is our original adaptation for this type of bone defect.^[10]

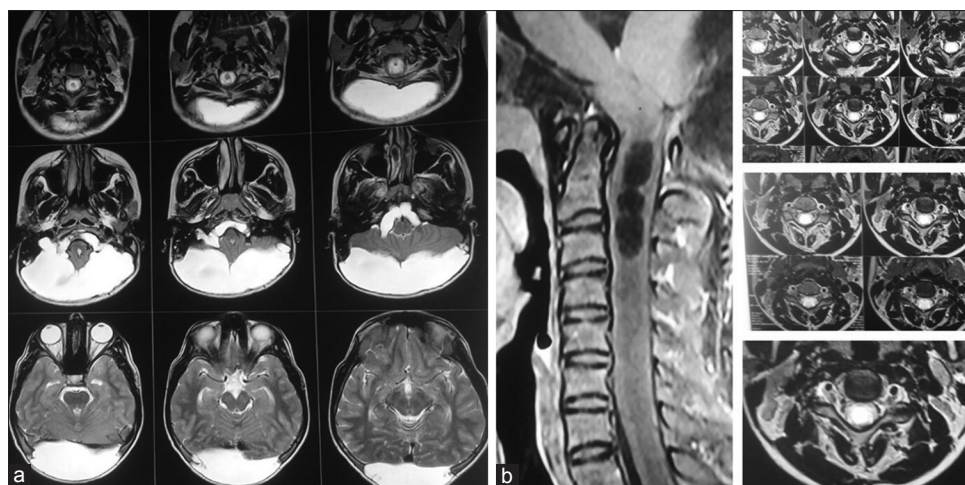


Figure 3: (a) Axial T2-weighted magnetic resonance images show the intradiploic cyst. Note that the signal intensity of the cyst is similar to that of the cerebrospinal fluid. (b) Sagittal T1-weighted and axial T2-weighted magnetic resonance images of cervical spine show the presence of syringomyelia extending from craniocervical junction to C3 vertebral body level

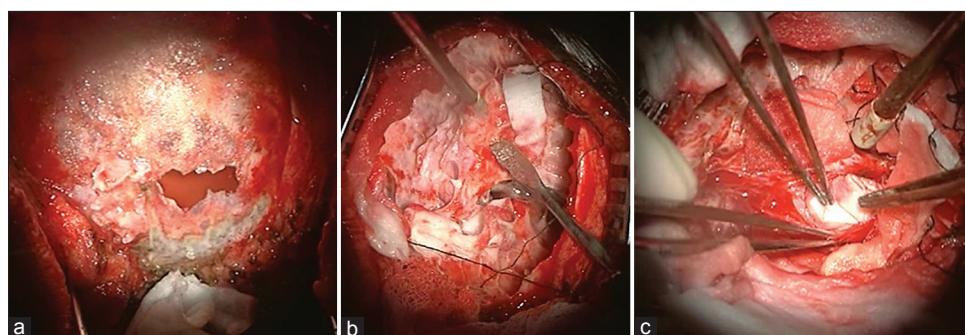


Figure 4: (a) Intraoperative photograph showing extremely thinned out outer table of the occipital bone along with evidence of cerebrospinal fluid within the intradiploic space. (b) Intraoperative photograph showing extremely thinned out inner table of the occipital bone (thinned outer table excised). (c) Small linear defect of 1 cm × 5 mm was visualized in the inner table along with underlying dural defect through which cerebrospinal fluid was coming into the intradiploic space

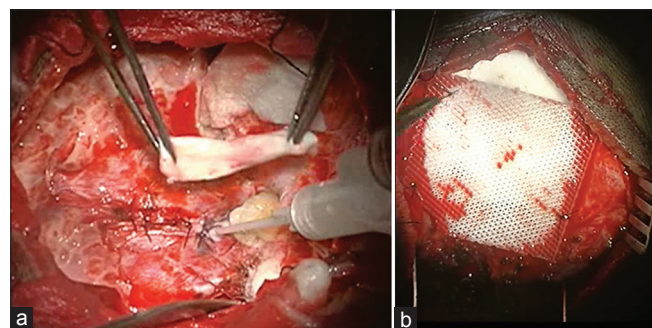


Figure 5: (a) Watertight repair of the dural defect was done using pericranium and then reinforced with fibrin glue. (b) Reconstruction of defect in the outer table was done using Osteomes

To our knowledge, this is only the seventh reported case of a large GSF of the posterior fossa which was intradiploic in the location.^[2,3,5-8] It demonstrates that a GSF may reach a considerable size, and although uncommon, intradiploic development and occipital localization of GSF are also possible.

Our case is unique from previously reported cases in regard to the fact that he had cervical syringomyelia,

which is highly unusual. We postulate that syrinx might be secondary to altered CSF dynamics at CVJ due to posttraumatic adhesions.

Informed consent

Written informed consent was taken from the parent for the publication of data in this journal.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

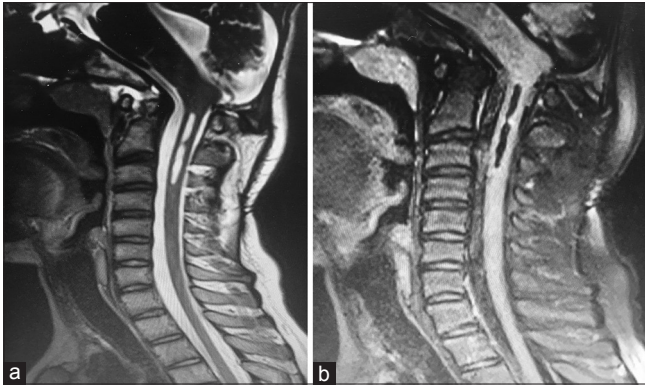


Figure 6: (a and b) Magnetic resonance imaging cervical spine done at 6-month follow-up showed a significant reduction in the size of syrinx

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