

## Spontaneous frontal intradiploic meningoencephalocele

Matthew James McPheeters, Daraspreet S. Kainth, Cornelius H. Lam

Department of Neurosurgery, University of Minnesota, Minneapolis, Minnesota 55455, USA

E-mail: \*Matthew James McPheeters - [mcph0066@umn.edu](mailto:mcph0066@umn.edu); Daraspreet S. Kainth - [kain0027@umn.edu](mailto:kain0027@umn.edu); Cornelius H. Lam - [lamxx023@umn.edu](mailto:lamxx023@umn.edu)

\*Corresponding author

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### Abstract

**Background:** Since 1976, 10 cases of intradiploic encephaloceles have been reported in the literature. This case is the first report of a spontaneous intradiploic meningoencephalocele of the frontal bone hypothesized to be secondary to distant head trauma.

**Case Description:** A 60-year-old female with a history of multiple traumatic head injuries as a child presenting with new onset generalized tonic-clonic seizures. Work-up revealed a right frontal epileptic focus. Imaging showed a right frontal intradiploic lesion. The patient underwent surgical resection, which during exploration was found to be an intradiploic encephalocele. She had an uneventful postoperative course with a resolution of seizures.

**Conclusions:** The authors hypothesize that the rare nature of posttraumatic frontal intradiploic encephaloceles is due to the increased thickness of the frontal bone compared to the parietal bone.

**Key Words:** Intradiploic, meningoencephalocele, trauma

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### INTRODUCTION

In 1976, Kosnik *et al.* published the first report of an intradiploic encephalocele.<sup>[6]</sup> The patient was a 57-year-old man who presented with generalized seizures and expressive aphasia.<sup>[6]</sup> Work-up revealed a left parietal lytic bone lesion on skull films, and an electroencephalogram (EEG) showed moderate slowing at 4 Hz with spike activity in the region underlying the skull lesion.<sup>[6]</sup> The patient underwent surgical resection of the encephalocele with a postoperative resolution of seizures.<sup>[6]</sup> Since this time, nine additional intradiploic encephaloceles have been reported in the literature. Eight of these cases were encephaloceles of the parietal bone, and the ninth was an iatrogenic encephalocele of the frontal bone.<sup>[1,3,4,7-9,12,13,15]</sup> We report the first spontaneous intradiploic meningoencephalocele of the frontal bone, which we hypothesize to be secondary to trauma from the distant past.

### CASE REPORT

A 60-year-old female has a history of multiple head traumas as a young adult, without documented skull

fracture, resulting from multiple falls while horseback riding. Her past history includes type two diabetes controlled with lifestyle modification and breast cancer in 2002 treated with surgical resection (2002) and chemoradiation (2003). She first presented with generalized tonic-clonic seizure. Her physical exam was notable for a palpable hard protuberance on her right frontal calvarium. An EEG showed intermittent low-amplitude polymorphic delta slowing over the right anterior head region, without clear epileptiform discharges, and she was treated with levetiracetam for seizure prophylaxis. Her neurological exam remained normal, and she did not have any subsequent seizures.

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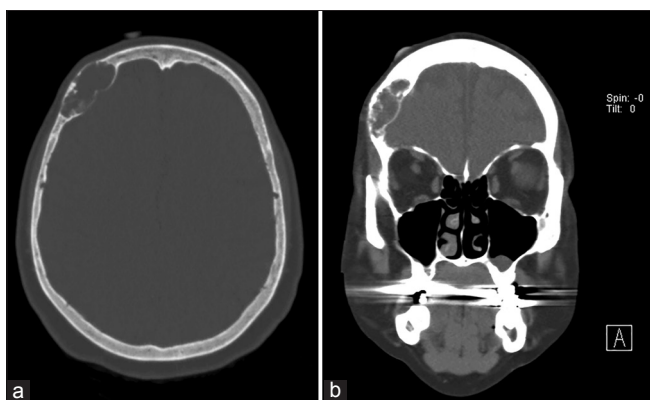
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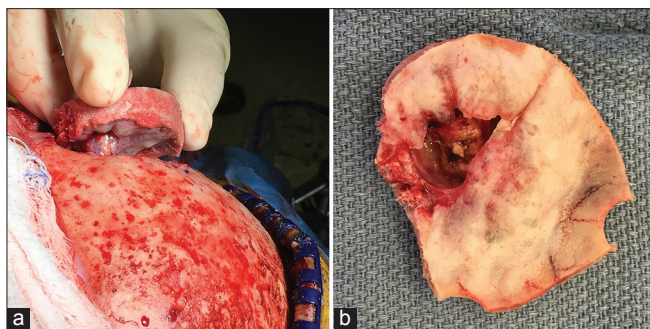
Imaging studies of her head were performed. The computed tomography (CT) of her head demonstrated a 3.7 cm × 3.4 cm × 1.5 cm lytic lesion of the right frontal calvarial bone with the erosion of the inner table [Figure 1]. The magnetic resonance imaging (MRI) of her brain suggested herniation of the right frontal lobe parenchyma into the bony defect [Figure 2].

### Operation

Due to the history of seizure activity and the expansile nature of the bony mass, the patient underwent surgical resection. A right frontotemporal craniotomy for resection of the right frontal calvarial lesion was performed. Upon inspection of the skull prior to the craniotomy, the lesion was evident by marked cystic thinning of the frontal bone. As the bone flap was elevated, brain tissue was noted to have herniated into the calvarial lesion [Figure 3a]. Bipolar electrocautery was used to coagulate and divide the herniated portion of the brain from the right frontal lobe, and the specimen was sent for frozen and permanent pathology. The bone flap was inspected on the surgical table [Figure 3b], and the inner table was removed prior to the replacement of the bone flap and closure.



**Figure 1:** (a) Axial computed tomography head demonstrating erosion of the inner table of the right frontal bone. (b) Coronal computed tomography head demonstrating erosion of the inner table of the right frontal bone



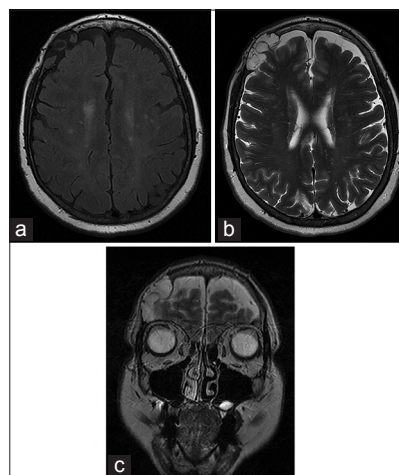
**Figure 3:** (a) Intraoperative photograph of meningoencephalocele herniating through the inner calvarial table. (b) Intraoperative photograph of frontal bone lesion

### Pathology

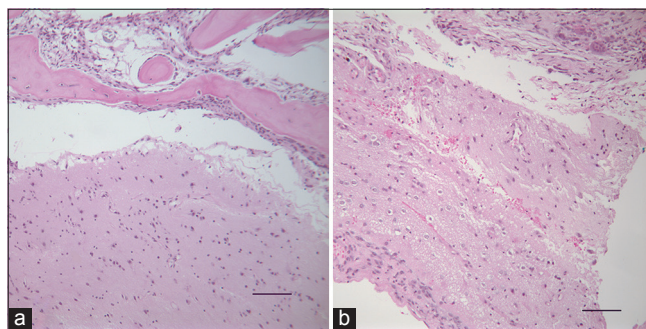
Frozen and permanent sections were taken of both the herniated brain tissue and bone lesion. The frozen sections demonstrated no definitive pathology with herniated cerebral cortex as well as a bone with fibrous tissue. The permanent sections, stained with hematoxylin and eosin [Figure 4], confirmed the frozen findings and also demonstrated gliosis of the cerebral cortex with thickening of the leptomeninges within the defect in the skull.

### DISCUSSION

Herniation of cerebral contents into the intradiploic space is an uncommon clinical scenario. Case reports have demonstrated multiple different etiologies including meningoencephaloceles, as in the above case, as well as giant arachnoid granulations, epidermoid cysts, and arachnoid cysts.<sup>[2,3,5,14]</sup> A cephalocele is



**Figure 2:** (a) Axial T1-weighted magnetic resonance imaging with heterogeneous hypointensity of the right frontal lobe lesion. (b) Axial T2-weighted magnetic resonance imaging with heterogeneous hyperintensity of right frontal lobe lesion. (c) Coronal T2-weighted magnetic resonance imaging with heterogeneous hyperintensity of right frontal lobe lesion



**Figure 4:** (a) Cerebral cortex with a bone, that is, intermingled with arachnoidal cells; H and E (scale bar = 100 μm). (b) Gliotic cerebral cortex with thickened leptomeninges; H and E (scale bar = 100 μm)

a herniation of intracranial contents through a bony or dural defect.<sup>[7]</sup> They are classified both by location (e.g., frontal, parietal, and occipital) and the herniated contents: meninges (meningeal) or meninges and parenchyma (meningoencephaloceles). Most commonly, cephaloceles are congenital defects occurring in the midline secondary to improper closure of the neural tube. Acquired cephaloceles occurring during adulthood have been attributed many etiologies, including: Infection, trauma, surgery, and tumors.<sup>[3,7]</sup> When there is no easily identifiable cause, they are known as spontaneous cephaloceles, and most commonly occur in the cranial sutures.<sup>[3]</sup> In this case, the spontaneous meningoencephalocele is hypothesized to have occurred secondary to a skull fracture in the distant past, although no clear documentation of such is found.

Intradiploic meningoencephaloceles are the result of a defect of the inner table of the calvarium and subsequent herniation of the meninges and cerebral parenchyma into the intradiploic space. Of the ten previously reported cases, nine presented within the parietal bone, and the tenth occurred within the frontal bone.<sup>[1,3,4,7-9,12,13,15]</sup> It is important to note that the etiology of the single case within the frontal bone was iatrogenic, occurring after an accidental tear of the dura during a craniostomy repair.<sup>[9]</sup> The presentation of intradiploic meningoencephaloceles tends to be associated with the location of the lesion and includes a headache, focal weakness, and seizures.<sup>[3,8]</sup>

Imaging studies aid the diagnosis of intradiploic encephaloceles. On CT, there is an erosion of the inner calvarial table, and on MRI the herniated parenchyma will appear hypointense on T1-weighted images and hyperintense on T2-weighted images.<sup>[3]</sup> Diagnosis is confirmed with a tissue sample demonstrating cerebral parenchyma, often with evidence of gliosis, and no evidence of other pathologies.<sup>[3]</sup> The clinical presentation, radiographic, and pathologic findings of the patient in this report are consistent with an intradiploic meningoencephalocele. In fact many of the findings including, the patient's age, presenting symptoms, and clinical course are very similar to report by Kosnik *et al.* from 1976.<sup>[6]</sup> However, two distinctions make this case notable. First, the patient's history of multiple traumatic head injuries and second the frontal location of the meningoencephalocele.

Of the 10 previously reported cases of intradiploic encephaloceles, only one was associated with head trauma. This case, reported by Patil and Etemadrezai occurred in a 61-year-old male who hit his head on a garage door approximately 1-year prior to presenting with a persistent lump on his head and no neurologic symptoms.<sup>[12]</sup> In this report, the authors compared the intradiploic meningoencephalocele to other posttraumatic

intradiploic cystic lesions, including arachnoidal cysts, leptomeningeal cysts, and cerebrospinal fluid (CSF) fistula.<sup>[12]</sup> The authors hypothesized that posttraumatic intradiploic meningoencephaloceles are a rare adult variant of growing skull fractures, where low-velocity blunt trauma only fractures the inner table, as opposed to a growing skull fracture in which both the inner and outer tables are disrupted.<sup>[11,12]</sup> The fracture of the inner table results in a dural tear; as the skull recoils, a negative pressure draws the underlying arachnoid and parenchyma into the intradiploic space.<sup>[12]</sup> In addition, posttraumatic intradiploic encephaloceles and growing skull fractures differ in the extent of damage to the underlying parenchyma. Growing skull fractures usually result in secondary and often severe damage to the underlying parenchyma due to the underlying mechanism of rapid brain growth and CSF pulsation in infants.<sup>[11]</sup> However, in the previously reported posttraumatic intradiploic encephalocele as well as in the present case there was minimal damage to the underlying parenchyma.<sup>[12]</sup> This is likely due to the head injuries occurring after the period of rapid brain growth signified by the closure of the cranial sutures, and thus limiting the mechanism of secondary damage to the underlying parenchyma.

The frontal location is an additional distinction in this patient's meningoencephalocele. This case is the first reported spontaneous intradiploic encephalocele of the frontal bone. It is unclear why frontal intradiploic encephaloceles are less common than those of the parietal bone. A possible explanation is the difference in the average thickness of the two bones. The parietal bone is on average thinner than the frontal bone,<sup>[10]</sup> and is, therefore, likely to be more susceptible to developing posttraumatic intradiploic encephaloceles. However, this does not explain the increased frequency of spontaneous parietal intradiploic encephaloceles. Given that the trauma in posttraumatic intradiploic encephaloceles may occur 1 or more years prior to presentation without neurological deficit, many seemingly spontaneous intradiploic encephaloceles may be posttraumatic with the traumatic event not recalled or believed to be relevant by the patient. If trauma is the underlying cause of many spontaneous intradiploic encephaloceles, then this hypothesis would explain the increased incidence of intradiploic encephaloceles in the parietal bone as compared to the frontal bone.

## CONCLUSIONS

Posttraumatic intradiploic meningoencephaloceles remain a rare clinical entity. They are most likely a variant form of growing skull fractures and may be the underlying etiology of many spontaneous intradiploic encephaloceles. This case represents the first reported spontaneous intradiploic meningoencephalocele of the frontal bone

which is believed to have occurred secondary to head trauma in the distant past.

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### Conflicts of interest

There are no conflicts of interest.

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