

# Intradiploic Arachnoid Cyst with Meningothelial Hyperplasia: A Case Report

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We report the case of a 40-year-old man presenting with focal headache and a bulge at the right parietal bone, diagnosed as an intradiploic arachnoid cyst. The cyst wall included “meningothelial hyperplasia,” which is a rare finding. While over 40 cases of intradiploic arachnoid cysts have been reported to date, meningothelial hyperplasia in an intradiploic arachnoid cyst does not appear to have been reported. We also discuss the pathological findings of arachnoid cysts with meningothelial hyperplasia and mechanisms of enlargement of the arachnoid cyst.

**Keywords:** intradiploic arachnoid cyst; intraosseous arachnoid cyst; meningothelial hyperplasia; time-SLIP

## Introduction

Arachnoid cysts are benign abnormalities that usually exist in the intracranial regions, and rarely in the intradiploic region. The term ‘intradiploic arachnoid cyst’ was coined by Weinand et al. in 1989,<sup>1</sup> and has been used in various reports.<sup>2,3</sup> Various other names have been used to describe intradiploic cystic lesions in the literature, including intraosseous arachnoid cysts, intraosseous leptomeningeal cyst, traumatic/posttraumatic arachnoid cyst and intradiploic pseudomeningocele.<sup>4–6</sup> The definitions for these terms are overlapping and confused, with the exception of obvious pathological diagnoses including epidermoid cyst, dermoid cyst and meningoencephalocele.<sup>7</sup> Intradiploic cerebrospinal fluid (CSF) fistula is another cystic lesion, but usually contains a communication with the CSF space with no arachnoid membrane in the diploic space.<sup>8,9</sup> In the present case report, we use ‘intradiploic arachnoid cyst’ to mean an intradiploic (usually obstructive) cystic lesion covered with pathologically diagnosed ‘arachnoid membrane’. Over 40 cases of intradiploic arachnoid cyst and similar cases have been reported to date.<sup>1–6,10–12</sup>

We report herein a rare case involving an intradiploic arachnoid cyst with meningothelial hyperplasia. We also discuss the pathological findings in this case and mechanisms of enlargement of the arachnoid cyst.

## Case Presentation

A 40-year-old man with no previous medical history presented with a 3-year history of gradually exacerbating headache and a hard bulge in the right parietal region. The headache comprised focal pain above the bulge, with a visual analogue scale of 3 out of 10. The bulge was approximately 4 cm in diameter. He denied any history of head trauma, excluding playing rugby in his high school days. Apart from the bulge, no other positive findings were found in physical and neurological examinations. Laboratory examinations likewise revealed no abnormalities. Computed tomography (CT) of the head showed a low-density area at the parietal intradiploic lesion where the inner and the outer tables were extremely thin with defective parts (Fig. 1A). Magnetic resonance imaging (MRI) showed the contents of the lesion as hypointense on T<sub>1</sub>-weighted imaging (T<sub>1</sub>WI), hyperintense on T<sub>2</sub>-weighted imaging (T<sub>2</sub>WI) similar to the CSF, and hypo- to isointense on diffusion-weighted imaging (DWI) (Fig. 1B–D). No enhancement with gadolinium (Gd) was evident. No CSF flow was observable in intradiploic fluid on time-SLIP examinations using MRI.<sup>13</sup>

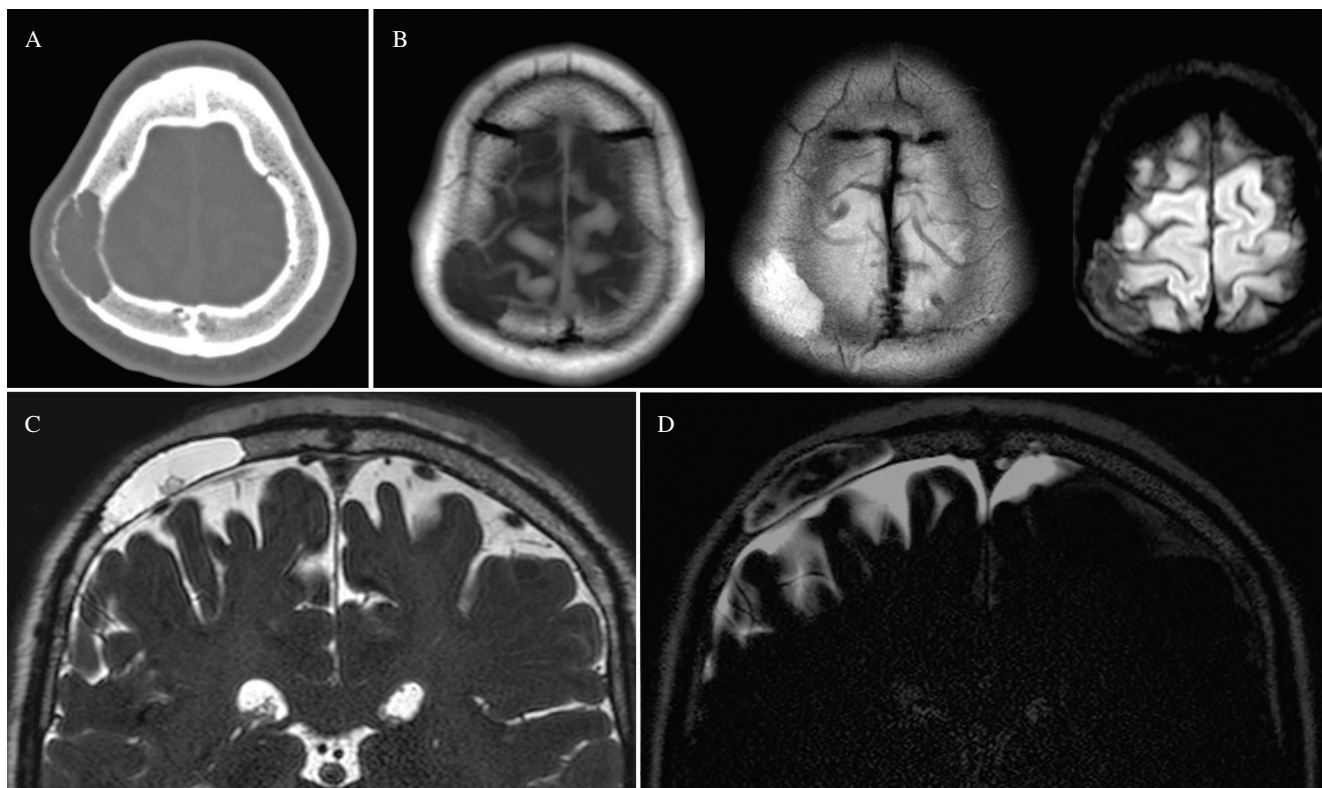
Surgery was performed to prevent cranial fracture. After the skin incision, the thinning outer table of the parietal skull bone with defective parts was revealed (Fig. 2A–F). After removing the outer table, the inner table was revealed accompanied by a bone defect 6 mm in diameter and an arachnoid granule-like protrusion. We extended the hole to reveal the dural layer and found the protrusion arising from under the dura mater through a hole. After eliminating the main part of the protrusion, no CSF leakage was seen with the Valsalva maneuver. The defect in the dura mater was closed with periosteum in a water-tight manner and the bone defect was filled with hydroxyapatite. The headache and bulge of the patient were resolved postoperatively. Neither side effects nor relapse have been reported at two month follow up.

Examination of the liquid contents of the cyst showed a high concentration of protein (338 mg/dl) compared to normal control data (10–40 mg/dl) for CSF, while other results were slightly apart from control data (sugar, 84 mg/dl (normal, 50–75 mg/dl); Cl, 118 mEq/l (normal, 120–125 mEq/l); cell count, 62 in 3 fields; mononuclear/polynuclear cell ratio, 44/18).

Histopathological examination revealed that the cyst wall comprised meningothelial cells in a single layer leading to the diagnosis of arachnoid cyst (Fig. 2G–K). The protrusion

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**Fig. 1** (A) CT showing extension of the intradiploic lesion into the right parietal bone. The outer and inner tables are thin with partial bone defects. (B) Axial MRI. Left, axial T<sub>1</sub>-weighted imaging; middle, axial T<sub>2</sub>-weighted imaging; right, diffusion-weighted imaging. (C) Coronal thin-slice drive-T<sub>2</sub>-weighted imaging. (D) Time-spatial labeling inversion pulse examination.

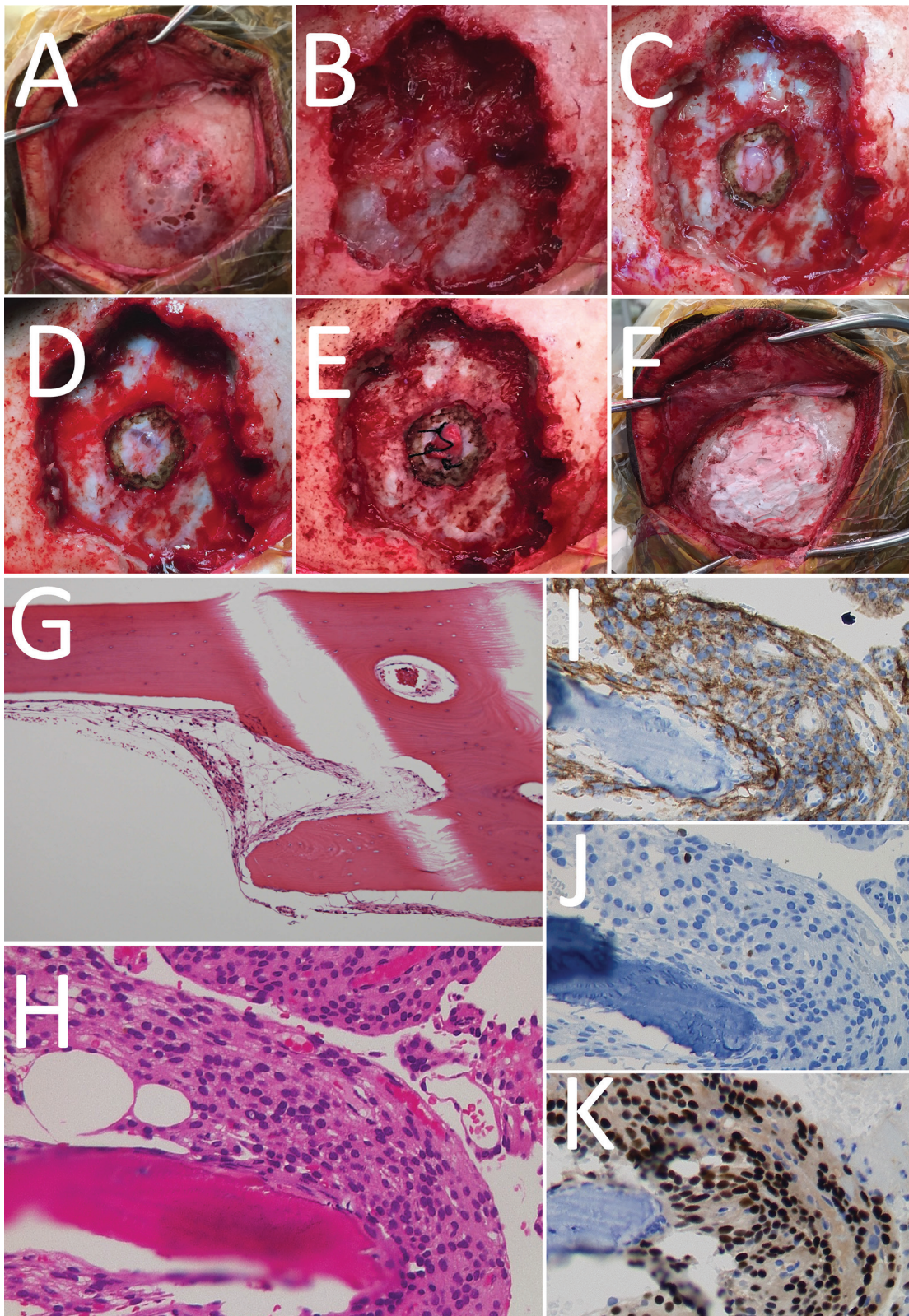
consisted of a thick layer of meningotheial cells with highly dense nuclei. Immunohistochemical examination revealed the protrusion was positive for epithelial membrane antigen (EMA) and progesterone receptor (PR), but negative for glial fibrillary acidic protein. Ki67 labeling index was less than 1%. These features led to a diagnosis of meningotheial hyperplasia.

### Discussion

The causes of intradiploic cystic formation including arachnoid cyst are considered to be posttraumatic or non-traumatic including congenital.<sup>1-6,10-12</sup> Posttraumatic causes can be not only traumatic, but also iatrogenic due to cranial surgery.<sup>9</sup> Non-traumatic intradiploic arachnoid cyst is characterized by multiple, parasagittal (commonly in the occipital region), well-demarcated osteolytic lesion, while the etiology remains unclear.<sup>12</sup> In our case, the patient's experience as a rugby player could have been involved in traumatic etiology, as in previously reported cases,<sup>8</sup> but the possibility of non-traumatic etiology cannot be denied. Due to such causes a defect is developed (or initially exists) in the inner table of the skull bone and the dura mater. With the pulsation of the CSF and the pressure gradient between the inner and outer tables, the arachnoid membrane herniates from the defect into the intradiploic area and gradually enlarges, leading to cyst formation. The continual pressure from the cyst wall on the outer table may cause thinning of the bone and erosion. The fragile outer table gradually enlarges, which would be

recognized as a bulge.<sup>2,11</sup> Furthermore, our case showed the possibility of a specific one-way valve mechanism in enlargement of the cyst (Fig. 3). Simultaneous with the formation of the intradiploic cyst, meningotheial hyperplasia also formed at the entrance of the arachnoid cyst, probably resulting from physical stimulation to regular arachnoid membrane or small arachnoid granulation. The hyperplasia gradually narrowed the entrance of the enlarged arachnoid cyst, allowing CSF into the cyst while preventing the outward flow through a one-way valve-like mechanism, before finally obstructing the entrance. The worsening headache reported by the patient could have been a sign of increasing pressure within the cyst, supporting the one-way valve theory. Furthermore, the fluid contents of the cyst resembled CSF, with the exception of the high concentration of protein. These findings could suggest that the cyst was a closed space.

Histopathological results in the present case revealed an interesting finding of meningotheial hyperplasia. The cause of meningotheial hyperplasia is thought to be reactive processes such as inflammation, formation of granulation tissue and fibrosis.<sup>14</sup> In that study, meningotheial hyperplasia was defined as a "reactive process characterized by a proliferation of arachnoid cap cells that is often non-invasive, multicentric, and at least focally reaches a thickness of 10 or more cell layers".<sup>14</sup> Meningotheial hyperplasia shows some overlap of histopathological features with meningioma and normal arachnoid tissue including arachnoid granulation. Arachnoid granulation is typically consisted of small nodule



**Fig. 2** Intraoperative photographs. (A) The outer table is thin with partial defects. (B) The inner table and arachnoid granule-like protrusion. (C–F) Photographs after eliminating the inner table around the protrusion (C), after removing the protrusion (D), after duraplasty with periosteum (E), and after cranioplasty with hydroxyapatite (F). Photomicrograph of surgical specimens. (G) The arachnoid tissue undercoats the skull in the outer table with cyst wall, (hematoxylin and eosin (HE) stain,  $\times 100$ ). (H) In the protrusion, meningothelial cells formed more than 10 layers and the nuclei are highly dense, but cells show no abnormal structures HE stain,  $\times 200$ ). (I–K) Immunohistochemistry of the protruding specimen (upper, EMA stain; middle, Ki-67 stain; lower, PR stain,  $\times 200$ ).

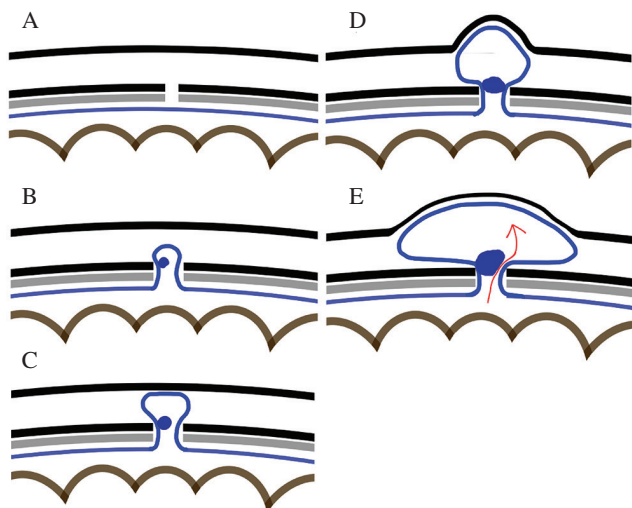


Fig. 3 Hypothesis for formation of the intradiploic arachnoid cyst in the present case. Layers comprise the outer table (black), inner table (black), dura mater (gray), arachnoid (blue), and brain (brown). (A) The inner table and dura mater develop a defect through trauma or non-traumatic cause. (B) The arachnoid herniates into the intradiploic area. Part of the arachnoid membrane starts to proliferate (meningothelial hyperplasia) due to reactive processes. (C) Arachnoid membrane enlarges due to pulsation of the CSF and exerts outward pressure on the outer table, resulting in enlargement. (D) The meningothelial hyperplasia narrows the entrance to the cyst. (E) The hyperplastic lesion works as a one-way valve and finally occludes the entrance.

formed from arachnoid cells that have a tiny or flat nucleus. The protrusion in our case consisted of multilayered arachnoid cells with nucleus showing very fine and dense chromatin. These findings represent that the cells possess higher proliferative ability than regular arachnoid cells/granulation. Distinguishing between meningothelial hyperplasia and small meningioma clinically is particularly difficult since both can show arrangements of whorls and/or psammoma bodies, while only small meningioma shows dural invasion. The protrusion in the present case was compatible with the above criteria for meningothelial hyperplasia. In addition, the lack of dural invasion and very low Ki-67 labeling index of the cells supported the diagnosis of meningothelial hyperplasia, although the original source of these cells could be regular arachnoid membrane or small arachnoid granulation.

We should preoperatively differentiate an arachnoid cyst from osteolytic lesions including epidermoid cyst and meningocele. DWI is a useful modality to differentiate an arachnoid cyst from epidermoid cyst, since the former shows signal hypointensity, whereas the latter shows signal hyperintensity.<sup>15)</sup> Both arachnoid cyst and meningocele show signal

hypointensity on T<sub>1</sub>WI, signal hyperintensity on T<sub>2</sub>WI, and signal hypointensity on DWI. In our case, the time-spatial labeling inversion pulse (SLIP) sequence on MRI<sup>13)</sup> to observe CSF flow and identify the communication between the cyst and arachnoid space matched the operative findings. However, it is uncertain whether this sequence is truly available for detecting very small fistula in diploic lesion. Further studies of such lesions are needed to clarify this issue.

In conclusion, we have reported a rare case of an intradiploic arachnoid cyst containing meningothelial hyperplasia that might have acted as a one-way valve.

## Disclosure Statements

The authors have no conflict of interest.

## References

- Weinand ME, Rengachary SS, McGregor DH, Watanabe I: Intradiploic arachnoid cysts: report of two cases. *J Neurosurg* 70: 954–958, 1989
- Martinez-Lage JF, Martinez Pérez M, Domingo R, Poza M: Posttraumatic intradiploic arachnoid cyst of the posterior fossa. *Childs Nerv Syst* 13: 293–296, 1997
- Verma SK, Satyarthee GD, Sharma BS: Giant intradiploic arachnoid cyst for 13 years. *J Pediatr Neurosci* 9: 139–141, 2014
- Kumar R, Chandra SP, Sharma BS: Giant intradiploic pseudomeningocele of occipital bone. case report. *J Neurosurg Pediatr* 9: 82–85, 2012
- Mahapatra AK, Tandon PN: Post-traumatic intradiploic pseudomeningocele in children. *Acta Neurochir (Wien)* 100: 120–126, 1989
- Menkü A, Koç RK, Tucer B, Akdemir H: Is skull fracture necessary for developing an intradiploic pseudomeningocele as a complication of head injury in adulthood? *Acta Neurochir (Wien)* 146: 623–627, 2004
- Patil AA, Etemadzezaie H: Posttraumatic intradiploic meningoencephalocoele Case report. *J Neurosurg* 84: 284–287, 1996
- D'Almeida AC, King RB: Intradiploic cerebrospinal fluid fistula. Report of two cases. *J Neurosurg* 54: 84–88, 1981
- Placantonakis DG, Lis E, Souweidane MM: Intradiploic cerebrospinal fluid fistulas of iatrogenic origin. Report of two cases. *J Neurosurg* 104 (5 Suppl): 356–359, 2006
- Asahi T, Endo S, Akai T, Takaba M, Takaku A: Nontraumatic convexity intradiploic arachnoid cyst. *Neurol Med Chir (Tokyo)* 38: 374–376, 1998
- Sartawi M, Schwartz FT, Fox JL: An unusual osteolytic lesion of the skull due to a traumatic arachnoid cyst. *Neuroradiology* 6: 180–181, 1973
- Hasegawa H, Bitoh S, Koshino K, Obashi J, Iwaisako K, Fukushima Y: Nontraumatic intradiploic arachnoid cysts: report of five cases. *Neurol Med Chir (Tokyo)* 32: 887–890, 1992
- Miyazaki M, Kanazawa H, Higashi M, Morohoshi Y, Bluml S, McComb JG: Visualization of cerebrospinal fluid movement with spin labeling at MR imaging: preliminary results in normal and pathophysiologic conditions. *Radiology* 249: 644–652, 2008
- Perry A, Lusus EA, Gutmann DH: Meningothelial hyperplasia: a detailed clinicopathologic, immunohistochemical and genetic study of 11 cases. *Brain Pathol* 15: 109–115, 2005
- Yamaguchi S, Hirohata T, Sumida M, Arita K, Kurisu K: Intradiploic arachnoid cyst identified by diffusion-weighted magnetic resonance imaging case report. *Neurol Med Chir (Tokyo)* 42: 137–139, 2002

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