Intradiploic occipital pseudomeningocele in a patient with remote history of surgical treatment of Chiari malformation

Case report

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An intradiploic CSF pseudocyst is a rare entity that has been described in association with trauma, as a sequela of untreated hydrocephalus, or occasionally as a congenital finding in older adults. The authors present the case of a woman with a remote history of a posterior fossa intradural procedure, in which she underwent Chiari malformation decompression, Silastic substitute–assisted duraplasty, and occipitocervical fusion; she presented 19 years later with recurrent symptoms of Chiari malformation. She was found to have an occipital intradiploic pseudomeningocele, arising within her dorsal occipitocervical fusion mass and resulting in dorsal hindbrain compression. She underwent a posterior fossa decompression and revision of her failed duraplasty, and she had a good recovery. This case demonstrates intradiploic CSF pseudomeningocele as a rare potential delayed complication of an intradural procedure for the treatment of Chiari malformation with occipitocervical fusion.

KEY WORDS • occipitocervical fusion • intradiploic pseudocyst • posterior fossa decompression • Chiari malformation • cervical

Intradiploic cerebrospinal fluid (CSF) pseudocyst or pseudomeningocele is an exceptional finding, with few cases described. Cases reported in the literature have been variably described as congenital intradiploic arachnoid cysts, as posttraumatic leptomeningeal cysts or pseudomeningoceles, or, less frequently, as intradiploic CSF pseudocyst presenting as a manifestation of unusual sequelae of hydrocephalus. Posttraumatic cases have been reported in children and adults and have been shown to be related to dural tears at the time of cranial trauma, with resultant intradiploic CSF cyst formation over the course of several years. In cases with no history of head trauma or cranial fracture, findings similar to those of idiopathic intradiploic arachnoid cysts have been described, and the lesions have been proposed to be congenital in nature. Even fewer cases have been described in which a CSF fistula occurs as a direct result of undertreated hydrocephalus. Intradiploic cysts (leptomeningeal cyst or arachnoid cyst) involving the cranial hemispheric convexities, the skull base, and the posterior fossa have been reported.1–5,11,13,17,18

We report a case of a delayed finding of a posterior fossa intradiploic CSF collection related to remote treatment of Chiari malformation with associated occipitocervical bony fusion in a patient originally treated as a young child.

Case Report

Presentation and History. A 21-year-old woman presented with numbness in the left forearm and headaches exacerbated by heavy lifting. Nineteen years previously, at the age of 2 years, she had undergone posterior fossa intradural decompression for the treatment of Chiari I malformation, duraplasty with a Silastic substitute, and occipitocervical fusion with an iliac crest bone graft.

Examination. The patient had normal cranial nerve function, including full extraocular motility without nystagmus. She exhibited weakness in hand grip bilaterally and diminished sensation to light touch in the left upper extremity. Proprioception was diminished in the left upper extremity. Deep tendon reflexes were diffusely diminished. The tandem Romberg test was positive, and the patient was noted to walk with small, mincing steps.

Imaging Findings. Lateral cervical spine plain radiographs demonstrated a honeycomb appearance to the dorsal fusion mass (Fig. 1). Computed tomography scans showed a dorsal bulbous fusion mass from the occiput to C-3, with cystic-appearing changes within the fusion mass.
Operation. An operation was undertaken with the patient prone in crown halo traction. The posterior dorsal fusion mass was encountered in the midline, and upon subperiosteal dissection, CSF was found to emanate from the left side of the fusion mass. The bone was markedly trabeculated with a honeycomb appearance of CSF-filled cells (Fig. 4). The trabeculated bone was removed using rongeurs and a diamond drill bit to create a 3 × 3–cm bony opening. Despite the honeycomb-appearing, fluid-filled consistency of the bone dorsally, a reasonably solid fusion had occurred laterally, and revision of the occipitocervical fusion was not required. A Silastic dural substitute from the patient’s prior Chiari malformation procedure was encountered with CSF egress around the dural substitute (Fig. 5 left). The Silastic dural substitute was removed and a cuff of dura preserved. Adhesions were observed between the tonsils in the midline, and there was obstruction of CSF outflow from the fourth ventricle (Fig. 5 right). The cerebellar tonsils were dissected carefully under the operative microscope, and the tonsils were shrunk using bipolar cautery at a low setting, preserving the pia membrane. Duraplasty was completed with fascia lata allograft sutured primarily to the native dura, followed by onlay of Durepair and DuraGen.

Postoperative Course. The patient did well, and by 3 months postoperatively she was already noted to have improvement in her cervical cord syringohydromyelia. She was last seen 4 years postoperatively. At that time, her syringohydromyelia had completely resolved (Fig. 3) and, her presenting symptoms having also resolved, she had returned to a fully functional lifestyle.

Discussion

To our knowledge, this is the first case report of an intradiploic CSF collection related to remote history of posterior fossa intradural procedure for Chiari malformation with application of a Silastic dural substitute and occipitocervical fusion. Other causes of intradiploic CSF pseudocysts have been described: traumatic intradiploic pseudomeningocele, iatrogenic intradiploic pseudomeningocele, nontraumatic/idiopathic/congenital intradiploic arachnoid cyst, and untreated hydrocephalus.

Traumatic Intradiploic Pseudomeningocele

Perhaps the most frequently described etiology of intradiploic CSF pseudocyst formation is the posttraumatic intradiploic pseudomeningocele. It is usually described as a very thin or even transparent outer cortical bone, trabeculated or fully eroded CSF-containing intradiploic spaces, and a very thin inner cortical table with varying degrees of erosion—often in association with the inciting fracture. A rare variant of growing skull fractures, this lesion is usually observed in the parietal or occipital region, although Mahapatra and Tandon described 1 frontal cyst and 1 orbital roof cyst (of a series of 8). Several cases have been described in the posterior fossa. It is thought that a fracture through the inner table of the skull and laceration of the underlying dura in the presence of focally increased pressure result in bone resorption and expansion of
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The diploic spaces. An intradiploic pseudomeningocele has also been suggested to possibly result from trauma, without underlying skull fracture. While a growing skull fracture more frequently presents as a soft scalp mass, an intradiploic pseudomeningocele may present as a firm, hard scalp mass. Frequently there is no associated neurological deficit, but neurological involvement may be dependent on location and extent of bone resorption. Kumar et al. described a case of a giant intradiploic pseudomeningocele involving the occipital bone that was a delayed result of trauma. This patient had a large posterior fossa lesion, and the dorsal compression had resulted in an extensive cervical cord syrinx that caused the patient to present with progressive spastic quadriparesis and loss of gag reflex.

Iatrogenic Intradiploic Pseudomeningocele

Iatrogenic intradiploic pseudomeningocele has also been described. In one case the lesion was related to an accidental dural tear during an operation for craniosynostosis. The child presented several years later with a hard, slowly progressing lesion in the frontal region and was found to have a CSF-containing intradiploic expansion of the frontal bone. A second case was reported in a child with a cerebellar medulloblastoma who underwent resection of the tumor at age 9 years, presented at age 16 with postural headaches, and was then found to have an occipital bone intradiploic CSF collection.

Nontraumatic/Idiopathic/Congenital Intradiploic Arachnoid Cyst

Nontraumatic or idiopathic intradiploic arachnoid cysts have been described as well. Typically, this lesion is seen in older or elderly adults and is a similar lesion to the traumatic intradiploic arachnoid cyst, but without history of trauma or evidence of fracture. A small dural defect with herniating arachnoid membrane is usually found on exploration. In some cases the cyst was reported as an incidental finding and in other cases the patient presented with headaches or localized pain. The pathogenesis of these lesions is thought to be developmental or congenital and analogous to that of congenital extradural spinal arachnoid cysts.

Sequela of Untreated Hydrocephalus

An intradiploic CSF pseudocyst has also been de-
scribed in association with untreated hydrocephalus—in the case of a patient with an unusual presentation of ventriculoperitoneal shunt malfunction. Sato et al. described the case of a 15-year-old with Dandy-Walker malformation, treated with a cystoperitoneal shunt connected to a ventriculoperitoneal shunt via a Y connector, who presented with an intradiploic cyst of the occipital bone.\(^7\)

On exploration, the occipital outer cortical bone was very thin, and an expanded intradiploic space containing CSF under high pressure was encountered. The proximal catheter of the cystoperitoneal shunt was found to be occluded and was revised.

In our case report, the patient had similar features to those of other cases of intradiploic CSF pseudocyst reported in the literature in that the intradiploic CSF collection and bony erosion were secondary to a dural defect with focally increased CSF pressure. In our patient’s case, the previously used Silastic dural substitute did not afford adequate dural closure over time and resulted in erosion and trabeculation of the intradiploic space of the bony mass, which had formed following dorsal occipitocervical fusion. With expansion of the bone, the patient began to experience symptoms of mass effect from dorsal compression, resulting in symptomatic presentation of Chiari malformation.

This case is distinct in not only the presentation of a patient with an intradiploic CSF collection and symptoms of Chiari malformation (only one other case is identified in the literature), but also in the etiology of presentation of this type of lesion, that is, as a sequela of treatment of Chiari I formation. Our case highlights the importance of achieving a watertight dural closure in the surgical treatment of Chiari malformation and in other posterior fossa surgical procedures in which CSF pressure may be focally or globally elevated. This case also alerts clinicians to an unusual cause of clinical deterioration in a patient treated for Chiari malformation in whom occipitocervical fusion has also been performed.

**Conclusion**

An intradiploic CSF pseudomeningocele is a rare potential delayed complication of surgical treatment of Chiari malformation and occipitocervical fusion.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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**References**


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