Transosseous cerebrospinal fluid fistula 14 years after Chiari decompression: presentation and management

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The authors report a unique case of a transosseous CSF fistula that was detected more than 10 years after treatment of a symptomatic Chiari I malformation. This lesion initially presented as an intraosseous cystic lesion involving the C-2 vertebra, which was found to communicate freely with the subarachnoid space through a tiny dural opening. Surgical management involved hemilaminectomy and repair of the dural defect followed by reinforcement of the bony defect with demineralized bone matrix. Following closure of the fistula, symptoms of elevated intracranial pressure developed, necessitating a ventriculoperitoneal shunt for CSF diversion.

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ACEREBROSPINAL fluid fistula is a unique etiology of an intraosseous spinal lesion. Acquired intraosseous CSF cysts in the cranial vault are usually associated with posttraumatic growing skull fractures.^{2,12} To the best of our knowledge, there have been no cases of spinal intraosseous CSF fistulas associated with a dural defect. Cystic lesions of the bone containing CSF of a nontraumatic origin are rare, apart from congenital lesions such as meningoceles. A literature search revealed reports of 5 such cases involving the cranial vault as well as a small series of 5 cases all involving the basiocciput.^{1,2,4,6,9,22} We report on a young boy with a spinal intraosseous cystic lesion containing CSF that manifested 14 years after a Chiari decompression.

Case Report

An 18-year-old male patient presented with a history of patchy numbness on his left thigh, which had resolved spontaneously several months before presentation; however, based on his prior history, he was referred for neurosurgical consultation. At the age of 4 years the patient had undergone posterior fossa craniectomy, C-1 laminectomy, and expansile duraplasty for clinical and radiological evidence of a Chiari I malformation that was associated with severe exertional headaches (Fig. 1 left). These symptoms

entirely resolved postoperatively, and the postoperative MRI study demonstrated adequate decompression of the Chiari malformation (Fig. 1 right). The patient remained asymptomatic for 14 years, although he had sustained several minor traumatic events in the interim, without apparent sequelae. On re-presentation there was no history of recent trauma, and the symptoms that had led him to seek neurosurgical reevaluation had entirely resolved. Findings from his neurological examination were normal.

An MRI study, which had been obtained by his primary care physician after his initial transient symptoms, revealed a honeycombed lesion within the right C-2 lamina, pedicle, and lateral vertebral body (Fig. 2) that was isointense to CSF on all imaging sequences. No compression of the thecal sac was apparent, and the Chiari malformation appeared well decompressed, without an obvious pseudomeningocele. CT scanning of the cervical spine showed an irregular expansile lesion involving the C-2 lamina, pedicle, and lateral mass on the right side, with scalloping and thinning of both outer and inner tables (Fig. 3). CT angiography demonstrated no evidence of vertebral artery compression caused by the lesion. The differential diagnosis based on the imaging appearance was a primary bony tumor, such as a chordoma or aneurysmal bone cyst, versus intraosseous CSF accumulation at the caudal aspect of the prior Chiari decompression site. A CT-guided biopsy/

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FIG. 1. Left: Preoperative sagittal T1-weighted MR image demonstrating the patient's Chiari malformation. Right: Sagittal T1-weighted MR image obtained 3 months postoperatively, demonstrating the decompressed Chiari malformation.

aspiration was performed and yielded crystal-clear fluid, consistent with CSF; because of this appearance, myelographic contrast was administered via the biopsy needle. A 22-gauge spinal needle was used to access the lesion from a posterior lateral approach. Two milliliters of Isovue-M200 (Bracco Diagnostics Inc.) was injected under CT fluoroscopic guidance, which demonstrated free fluid

of contrast into the subarachnoid space, as well as an obvious fistula in the dorsal midline (Fig. 4).

Because of the expansile nature of the lesion, with significant bony erosion, a C-2 hemilaminectomy was performed to explore and repair the intraosseous fistula. During surgery, the outer table of the C-2 lamina was found to be paper thin, and the intraosseous space was filled with

FIG. 2. Axial T2-weighted MR image demonstrating a right-sided honey-combed cystic lesion of the C-2 vertebra with signal isointense to CSF.

FIG. 3. Axial CT scan without contrast demonstrating bony anatomy of right-sided cystic lesion with thinning of bone and expansion of C-2 lamina, pedicle, and lateral mass.

FIG. 4. Axial CT scan obtained during CT-guided needle aspiration and myelographic contrast injection, demonstrating successful needle cannulation of cyst with injection of contrast. Midline communication with the thecal sac is indicated by the arrow.

CSF without the presence of a discernible lining. The inner table had also been thinned out by the lesion. At the base of the lamina, a small elliptical dural opening was visualized, measuring about 4 mm long, in the midline, through which CSF was found gushing out (Fig. 5). This dural opening appeared mature with fibrotic margins and was closed primarily after excision of the thinned-out bone. A watertight closure was obtained. The bony cavity between the inner and outer tables of the C-2 vertebra was reinforced using demineralized bone matrix.

Approximately 1 week after discharge from the hospital, the patient returned to the emergency department with headache, nausea, vomiting, and neck pain. CT imaging revealed a small pseudomeningocele in the surgical cavity. A lumbar puncture was performed, demonstrating an opening pressure of 51 cm H₂O; analysis of the sample revealed unremarkable cell counts and chemistry and negative cultures. Following the lumbar puncture, the patient's symptoms were temporarily relieved. When the symptoms returned, a lumbar drain was placed to reduce the size of the pseudomeningocele and give symptomatic relief. Lumbar drainage was performed over the course of 4 days. We then attempted to reduce the lumbar drainage in an effort to discontinue it, but the patient became increasingly symptomatic; his symptoms were relieved only with increased CSF drainage. Repeat neuroimaging during this period failed to reveal any ventricular enlargement over baseline but did show reduction in the size of the cervical

FIG. 5. Intraoperative photograph demonstrating the midline dural defect (arrow). Figure is available in color online only.

pseudomeningocele. Because of the continued need to divert CSF, we elected to place a ventriculoperitoneal shunt on Day 7. Following the placement of the shunt (Medtronic Strata programmable valve set at 2.5), the patient was relieved of his symptoms and was discharged the following day. At 1-month follow-up the patient continued to be symptom free and doing well.

Discussion

The differential diagnosis for a bony cavitory lesion includes intraosseous epidermoid/dermoid cyst, aneurysmal bone cyst, lymphatic malformation, eosinophilic granuloma, and malignant conditions such as plasmacytoma, myeloma, osteogenic sarcoma, chordoma, or chondrosarcoma, among others.^{3,5,8,10,13,15,18,19,21} Eosinophilic granuloma and malignant neoplasms are osteolytic rather than expansile lesions and thus differ in appearance from the current case. Although dermoid and epidermoid tumors and lymphatic malformations are often expansile rather than lytic, these lesions are not typically isointense to CSF on diffusion-weighted MR images, and thus are also distinct from the present case.

The lesion described in this report seems to most closely resemble a posttraumatic leptomeningeal cyst, which has been described cranially in association with a "growing skull fracture."^{12,14} This phenomenon occurs almost exclusively in children, resulting from pulsatile pressure of the growing brain through a fracture line associated with an unrecognized dural laceration, driving arachnoid membrane and CSF into and, in many cases, through the diploic space of the skull. A similar process may rarely follow a craniotomy in which a persistent CSF fistula leads to bony erosion and gradual defect expansion. In contrast to a true congenital meningocele,⁴ such lesions are not lined by dura or arachnoid.

Although subarachnoid-pleural and subarachnoid-mediastinal fistulas^{7,16,17,20} have been noted in the spine after

trauma (either as a result of rupture of a known spinal meningocele or in association with significant spinal column disruption and concomitant dural laceration), the existence of an acquired intraosseous CSF fistula similar to our case has, to our knowledge, not been previously reported. A very recent report by Mahaney and Menezes¹¹ described an occipital intradiploic pseudomeningocele, arising within the dorsal occipitocervical fusion mass and resulting in dorsal hindbrain compression in a patient with a remote history of Chiari decompression. In our case, a prior post-operative MRI study after the Chiari decompression 14 years earlier had not shown the existence of either a pseudomeningocele or fistula, and the presumption is that the expansile lesion occurred in the intervening time period. The proximity of the dural defect to the caudal-most aspect of the prior Chiari decompression suggests that either a dural opening had developed below the prior duraplasty and enlarged over time or that a dural tear had occurred as a result of one of the minor traumas that the patient had experienced during childhood. The extensive bony remodeling of the inner and outer tables of the C-2 vertebra also suggests that gradually, pulsations of CSF from the subarachnoid space into the soft cancellous portion of the vertebra led to the dramatic honeycomb appearance that was observed.

The mechanism for this process remains conjectural, although the observation that the patient developed symptoms of increased intracranial pressure shortly after closure of the dural fistula suggests that persistence and expansion of this connection likely reflected that this connection was being used as an accessory CSF drainage pathway or buffer. Over its many years of development and expansion, this accessory pathway may have become an essential CSF drainage mechanism. Equally conjectural is the possibility that the patient had an underlying component of hydrocephalus as the basis for his symptomatic Chiari malformation in early childhood. The immediate resolution of his symptoms after the initial surgery at age 4 years and lack of any subsequent symptoms for more than a decade thereafter makes this possibility less likely but does not rule it out as a potential explanation for this unusual case.

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