



Case Report

Chiari malformation and spinal interdural cyst: A proposed association and review of the literature

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ABSTRACT

Background: Interdural cysts are rare meningeal cysts with an unclear etiology. They are often mistaken for other mass lesions, including arachnoid cysts and tumors. Correctly identifying and classifying these cysts, as well as how they have formed in individual patients, are crucial to providing effective treatment options for patients.

Case Description: We report a case of a patient with shunted idiopathic intracranial hypertension who developed a symptomatic Chiari malformation and was subsequently discovered to have a spinal interdural cyst. The Chiari malformation was likely due to intracranial hypotension secondary to lumbar cerebrospinal fluid (CSF) diversion. Once the shunt was removed, a spinal interdural cyst became clinically and radiographically evident, and the Chiari resolved, suggesting that both entities were effects of shared CSF flow dynamics.

Conclusion: This cyst likely originated due to the trauma from remote repeated lumbar punctures and lumboperitoneal shunt placement, allowing CSF to enter the interdural space after the catheter was removed.

Keywords: Chiari, Cyst, Interdural, Meningeal, Shunt

INTRODUCTION

Interdural cysts are rare entities within the classification of meningeal cysts for which the etiology is often unclear. They are often mistaken for other mass lesions, including arachnoid cysts and tumors. A new classification schema has been introduced, and it is imperative to correctly identify and classify these cysts to provide effective treatment options for patients. Interdural cysts can exert mass effect on the adjacent neural elements, and as they are filled with cerebrospinal fluid (CSF), it is hypothesized that they contribute to the overall flow dynamics of the CSF space.

CASE REPORT

We present a case of a 30-year-old woman with a history of multiple inpatient admissions and outpatient visits for headaches due to idiopathic intracranial hypertension status – postlumboperitoneal (LP) shunt placement with a programmable valve 5 years before at another institution. She presented with new nonpositional tussive headaches, without any deficits on physical examination. Her LP shunt valve (Strata II) was confirmed to be set at 2.0, stable from 3 months before, when the resistance was increased from 1.0. Magnetic resonance imaging (MRI) was performed, demonstrating a Chiari I malformation [Figure 1a] without any abnormalities

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in the cervical spine [Figure 1b]. She was treated with LP shunt removal and placement of a new ventriculoperitoneal (VP) shunt (Certas valve set to 5). She had improvement in her headaches, however, she returned 3 months later with symptoms of bilateral upper extremity paresthesia. On physical examination, she exhibited full strength without hyperreflexia; however, she did endorse bilateral sensory dysesthesia when light touch was applied to her upper extremities.

Computed tomography (CT) and MRI studies

CT of the head and shunt series showed baseline small ventricle caliber and no shunt system disconnection, respectively. Her shunt remained at a moderate resistance (Certas Plus set at 5). Cervical MRI revealed resolution of the Chiari malformation. However, there was now an intradural extramedullary lesion ventral to the spinal cord starting at C3 and extending to L3. The lesion was hyperintense on T2-weighted imaging [Figure 2a], hypointense on T1-weighted imaging [Figure 2b], and displaced the cord posteriorly

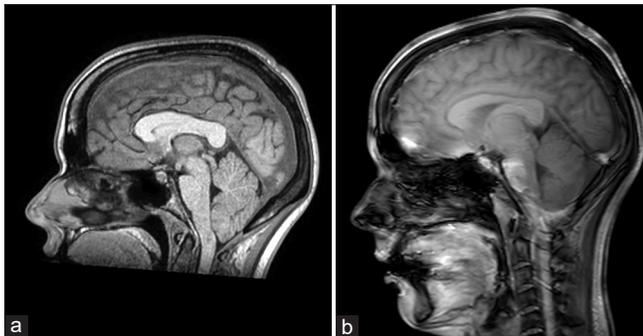


Figure 1: (a) Parasagittal T1-weighted MRI head demonstrating tonsillar herniation. (b) Sagittal MRI scout imaging demonstrating normal cervical spine, without any radiographic presence of an interdural cyst.



Figure 2: (a) Sagittal T2-weighted MRI cervical spine demonstrating high-intensity cystic cavity ventral to the spinal cord, isointense to cerebrospinal fluid, signified by black asterisks. (b) Sagittal T1-weighted MRI cervical spine demonstrating low-intensity cystic cavity ventral to the spinal cord, isointense to cerebrospinal fluid, signified by white asterisks.

[Figure 3]. The neurosurgery and radiology services concluded that this was an interdural cyst.

Surgery

Given her continued sensory symptoms, a decision was made to fenestrate the interdural cyst in the lower thoracic region.

A T11 laminectomy was performed, and the dura was opened just left of midline. After the arachnoid adhesions were released, a fluctuant dural surface was encountered anteriorly in the canal, with no other visible abnormality. The inner layer of the dura was incised and revealed a fluid-filled cavity, bounded by dura. This confirmed the presence of an interdural cyst [Figure 4]. Once adequate fenestration was accomplished and the cyst seemed decompressed, the wound was closed in usual fashion.

Postoperatively, the patient reported improvement in bilateral upper extremity tingling. At her 6-week follow-up appointment, she reported resolution of her paresthesias.

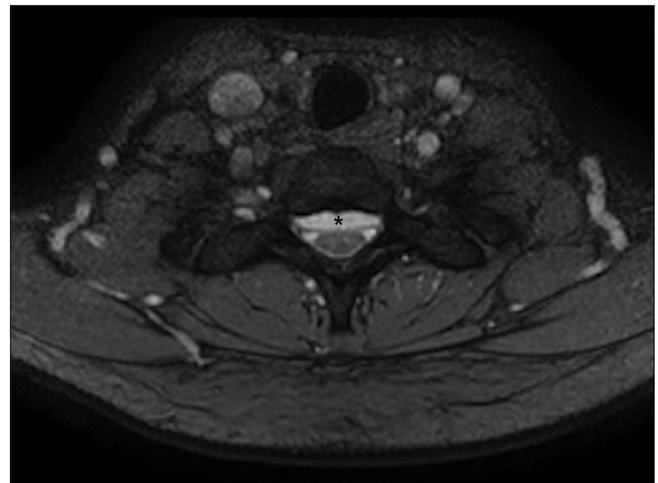


Figure 3: Axial T2-weighted MRI cervical spine demonstrating posterior displacement of the spinal cord due to mass effect exerted from ventral cystic lesion, signified by a black asterisk.

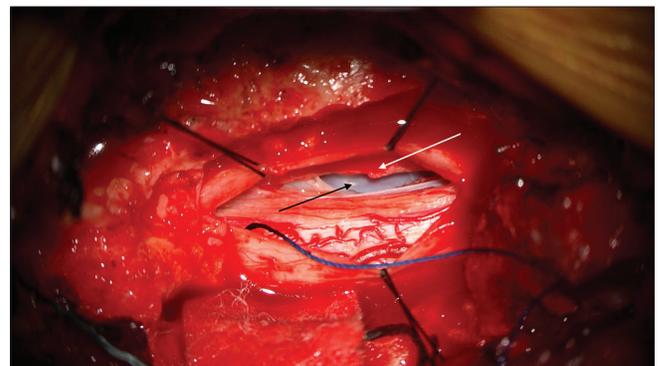


Figure 4: Intraoperative view of the cystic cavity with both superficial (white arrow) and deep (black arrow) dural leaflets present.

Head CT also demonstrated continued resolution of her Chiari malformation.

DISCUSSION

In 1988, based on a series of 22 cases, Nabors *et al.* attempted to describe an updated classification of spinal meningeal cysts, defined as diverticula filled with CSF, as extradural or intradural.^[8] Since then, there have been numerous reports describing the various types of cysts and their etiologies. Some have described congenital cysts as attributed to neural tube defects. There have also been reports of meningeal cysts developing secondary to inflammation, surgery, and other iatrogenic causes.^[9,10] However, the overwhelming majority of meningeal cysts reviewed in the literature were reported to develop after trauma.^[1,4,5]

Since the Nabors classification, there have been a number of case reports describing interdural cysts. Done *et al.* outlined a series of three patients who presented with cauda equina syndrome (two patients) and severe back and bilateral leg pain (one patient) and imaging findings of lumbosacral interdural cysts.^[2] Chen and Chen described a case of a cervical interdural cyst attributed to a traumatic C2 fracture.^[1] Lee *et al.* reported a case of the right leg pain and weakness associated with a lumbar interdural cyst.^[6] Sajjad *et al.* reported a case of a patient left T5 radicular pain associated with a thoracic interdural cyst.^[9] Details of these reports are found in [Table 1].

Due to an ever-growing recognition of this entity, new dural cyst classification systems have been proposed.^[5,9] In 2017, Klekamp described the largest case series of dural dissection (29 cases), a clinicopathologic entity analogous to interdural cysts. Nineteen of these patients lacked neural element herniation into the cyst. Trauma was the cause for

dural dissection in 15 patients, whereas the etiology for the remaining four could not be determined. Klekamp reported that mechanical compression from these dural dissections results in myelopathy and pain.^[5] He proposed a newer and more comprehensive classification system based on localization, communication with the subarachnoid space, etiology, involvement of nerve roots, and the spinal cord, and suggested management strategies for the various meningeal pathologies, including dural dissections.

A unique presentation

The case described here is unique given the discovery of a new Chiari malformation at initial presentation and subsequent diagnosis of the interdural cyst, implying possible relationship between the two entities. Before the patient's initial presentation, her LP shunt (Strata II valve) had been confirmed to be functioning. The setting was increased from 1.0 to 2.0 3 months before her initial presentation, thereby decreasing flow. Despite this, her imaging findings of Chiari malformation and clinical symptoms were consistent with intracranial hypotension. There are well-documented reports of intracranial hypotension resulting in an acquired Chiari I malformation.^[3,7]

Due to alterations of CSF flow dynamics following removal of the LP shunt with concurrent placement of a VP shunt and resolution of the Chiari malformation, we propose that a defect in the dura worsened, forming the interdural cyst. Subsequent expansion of this cyst exerted mass effect on her cervical spinal cord and resulted in her presenting with bilateral upper extremity paresthesia. We believe that the mechanism for a dural defect in this patient was trauma, whether from multiple lumbar punctures before shunting or placement of the lumbar shunt catheter itself.

Table 1: Review of interdural cysts in the literature.

Case	Clinical presentation	Etiology	Cyst location	Treatment	Recovery
Present case	Bilateral upper extremity paresthesia	Trauma	Cervical, thoracic, lumbar	Cyst fenestration	Complete
Sajjad <i>et al.</i> , 2015 ^[9]	Left T5 radicular pain	Unknown	Thoracic	Partial cyst wall excision and closure of defect	Complete
Lee <i>et al.</i> , 2010 ^[6]	Right L4 and L5 radicular pain and weakness	Unknown	Lumbar	Durotomy and drainage	Partial
Chen and Chen, 1996 ^[1]	Spastic left hemiparesis and numbness	Traumatic C2 fracture	Cervical	Durotomy and drainage	Partial
Done <i>et al.</i> , 1984 ^[2]					
Case 1	Cauda equina syndrome	Congenital	Lumbosacral	Cyst wall excision and closure of defect	Partial
Case 2	Cauda equina syndrome	Congenital	Lumbosacral	Cyst wall biopsy and closure of defect	Partial
Case 3	Back and bilateral lower extremity pain	Trauma	Thoracolumbar	Closure of defect	Complete

CONCLUSION

We report an unusual case of acquired Chiari malformation in a patient with an LP shunt who then presented with an extensive, and symptomatic, spinal interdural cyst after removal of the LP shunt, likely due to trauma from remote repeated lumbar punctures and LP shunt placement, allowing CSF to enter the interdural space after the catheter was removed; after fenestration of the cyst, the patient reported resolution in symptoms.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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