Case Report

Neurenteric cyst secondary to lumboperitoneal shunt

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ABSTRACT

Background: Neurenteric cysts are rare lesions that typically present in the upper thoracic and cervical spine and are occasionally found intracranially. The optimal treatment is gross total excision as subtotal/partial excisions are associated with high recurrence rates.

Case Description: For the past 10 years, a patient with pseudotumor cerebri required repeated lumboperitoneal (LP) shunt revisions. This resulted in multiple neuroenterogenous cysts occurring around the proximal LP subarachnoid shunt catheter, a finding likely attributable to retrograde flow from the peritoneal cavity.

Conclusion: Unlike ventriculoperitoneal (VP) shunts and LP shunts do not contain valves, making the retrograde passage of enterogenous cells possible when abdominal pressure exceeds lumbar subarachnoid pressure, especially in the morbidly obese patient.

Keywords: Enteroogenous cell, Neurenteric cyst, Pseudotumor cerebri, Morbid obesity, Idiopathic intracranial hypertension

INTRODUCTION

Neurenteric cysts, also known as enterogenous cysts, account for 0.3–1.3% of all spinal axis tumors. These rare lesions are formed during the 3rd week of development and are attributed to inappropriate partitioning of the embryonic notochord plate/endoderm. They are typically solitary and are commonly located in the upper thoracic or cervical spine, being rarely found intracranially. Complete resection of these intradural extramedullary lesions is feasible in 71–75% of cases and is essential to avoid high recurrence rates seen for subtotal/partial resections.

Here, we present a 42-year-old female with multiple enterogenous cell deposits in the lower thoracic spine found 22 years after the original insertion of a lumboperitoneal shunt.

CASE REPORT

Twenty-two years ago, a 42-year-old female presented with worsening vision/bilateral papilledema attributed to pseudotumor cerebri (idiopathic intracranial hypertension); a lumboperitoneal...
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(LP) shunt was placed and required revision 10 years later. Now, 22 years after the original surgery, this morbidly obese patient (body mass index of 52) newly presented with the 2-month onset of severe thoracic pain, bilateral leg weakness (grade 1/5), and a T1 sensory level.

MR studies

The thoracic MR demonstrated a hypointense lesion on the T1-weighted images that did not enhance with gadolinium. Notably, two well-defined, dorsal hyperintense lesions opposite the T9 and T11 levels flattened the spinal cord as best seen on the T2-weighted images [Figure 1a]. She underwent a T9–T12 laminoplasty for gross total excision of these lesions; at surgery, two cysts were found close to the subarachnoid shunt tip. They contained gelatinous watery fluid and a membrane that was slightly adherent to the pial surface. Postoperatively, she improved rapidly, and within 3 months, regained the ability to ambulate without assistance. The subsequent postoperative magnetic resonance imaging (MRI) scan confirmed complete lesional excision [Figure 1b].

Pathology

The gross pathological examination demonstrated multiple pieces of grey-white soft-tissue fragments measuring 2 × 1 × 0.2 cm. Microscopically, the specimen revealed a cyst lined by ciliated columnar epithelium [Figure 2a and b], The epithelium expressed CK7, WT-1 [Figure 3], and epithelial membrane antigen but not S100 protein and carcinoembryonic antigen. Hemosiderin pigment was focally present with underlying fibrous tissue and was most likely secondary to catheter induced trauma. The diagnosis of a neurenteric cyst was confirmed.

Subsequent gastric bypass surgery

The patient later underwent a bariatric gastric bypass. She was asymptomatic for the next year. Three years later, she has maintained her neurological baseline, for example, a high level of independent function.

DISCUSSION

Unlike VP shunts and LP shunts do not contain valves, making retroperitoneal passage of enteric cells possible when intra-abdominal pressure exceeds lumbar subarachnoid pressure. These enlarged enteric cysts, depending on their spinal location (e.g., cervical/thoracic/lumbar), are capable of causing significant compression leading to paraparesis. Lambert et al. previously established a relationship between morbid obesity and increased intra-abdominal pressure contributing to retrograde flow of enteric cells from the peritoneal cavity resulting in enterogenous mucin secreting cells located around a proximal LP shunt catheter. In Gallmann et al., multiple intraventricular fat deposits documented on MRI/computed tomography in a 25-year-old female were also attributed to retrograde flow from the abdominal fat occurring during multiple shunt revisions. Desai et al. reported the dissemination of enterogenous cells along any patent tract resulting in communicating...

Figure 1: (a) Preoperative mid-sagittal T2 magnetic resonance imaging spine showing two extradural intramedullary hyperintense lesions at the level of T9 and T11, (b) T2 mid-sagittal magnetic resonance imaging spine of the same level with complete excision of the lesions after surgery.

Figure 2: (a) H and E stained slide of cystic lesion (×100), (b) H and E stained section of the cyst wall lined by columnar ciliated epithelium (×400).

Figure 3: The cyst lining epithelium is immunoreactive for CK-7 (a, ×400) and WT-1 (b, ×100).
hydrocephalus (e.g. in a patient without a history of previous surgery).\[2\]

In the patient presented with morbid obesity and pseudotumor cerebri, the patient developed an enterogenous cyst at the proximal end of an LP shunt catheter at the T9 and T11 levels 22 years after having originally undergone LP shunt placement. Due to the marked cord compression, these lesions were appropriately excised, and the patient’s symptoms resolved.

Here, the enterogenous cysts were attributed to retrograde flow occurring when intra-abdominal pressure exceeded subarachnoid pressure in this morbidly obese patient. Future reduction in the intra-abdominal pressure with stringent weight loss, including, in this case, bariatric surgery should reduce the future risk of retrograde flow of enteric cells from the abdominal cavity to the subarachnoid space.

CONCLUSION

Retrograde flow of enteric cells from the abdominal cavity to subarachnoid space can occur and can be prevented with weight reduction which include in this case bariatric surgery.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Conflicts of interest

There are no conflicts of interest.

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