Iatrogenic cerebrospinal fluid leak and intracranial hypotension after gynecological surgery

Case report

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Perineural cysts are common lesions of the sacral spine. They have rarely been reported in a presacral location, leading to their misdiagnosis as a gynecological lesion. The authors report the second such case, in a patient undergoing fenestration of what was presumed to be a benign pelvic cyst, and the resultant high-flow CSF leak that occurred. They describe the clinical presentation and manifestations of intracranial hypotension, as well as the pertinent investigations. They also review the literature for the best management options for this condition.

Although they are uncommon, large perineural cysts should be included in the differential diagnosis when examining patients with a pelvic lesion. Appropriate imaging investigations should be performed to rule out a perineural cyst. The CSF leak that occurs from iatrogenic cyst fenestration may not respond to traditional first-line treatments for intracranial hypotension and may require early surgical intervention. The authors would recommend neurosurgical involvement prior to definitive treatment.

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Key Words • iatrogenic injury • complication • intracranial hypotension • review

Perineural cysts are common lesions most often arising from dorsal sacral nerve roots. These lesions have rarely been reported in a presacral location, leading to their misdiagnosis as a gynecological lesion. Recognition of the nature of this lesion is crucial in determining appropriate management. We report a case of a large presacral arachnoid cyst that was treated by marsupialization. The resultant high-volume subarachnoid pelvic CSF fistula caused severe symptomatic intracranial hypotension, necessitating surgery to repair the leak. It is only the second such case ever reported. A review of the literature and management options are presented.

Case Report

History and Examination. A 53-year-old woman with a history of neurofibromatosis Type 1 was referred to the neurosurgical service after presenting with protracted headaches. Her initial cranial imaging demonstrated bilateral subdural fluid collections, as well as pronounced cerebellar tonsillar herniation (Fig. 1). Her headaches had begun shortly after she underwent elective marsupialization of what was thought to be a benign pelvic wall cyst. Egress of copious clear fluid was reported at the time of surgery. Microscopic examination of the cyst wall revealed simple epithelial cells. Abdominal and pelvic imaging obtained after admission (in comparison with presurgical imaging) revealed a presacral arachnoid cyst arising from a sacral nerve root that after pelvic surgery had become small, irregularly shaped, and filled with hematoma (Fig. 2). Her clinical and radiographic findings supported a provisional diagnosis of symptomatic intracranial hypotension secondary to a subarachnoid-pelvic CSF fistula. Conservative management in the form of bed rest and placement of a lumbar drain was unsuccessful; after consultation with the general surgery service, operative repair was elected.

Operation. Under general anesthesia and with the assistance of a general surgeon, a transperitoneal approach was carried to the posterior wall of the peritoneal cavity. A copious quantity of clear, sanguineous fluid was identified. The marsupialized cyst was seen on the left posterior pelvic wall. A fistula spontaneously releasing moderate amounts of clear fluid was immediately seen. This fluid was found to be beta transferrin–positive, confirming it to be CSF. A pedicled flap of the omentum was mobilized and sutured in a purse-string fashion around the opening of the fistula. Approximately 5 ml of Tisseel (fibrinogen, thrombin, and aprotinin; Baxter) was then used to reinforce the mend. Immediately after opposition of the omentum, no further egress of CSF was noted. A Valsalva maneuver did not induce further egress of fluid.

Postoperative Course. Immediately after surgery the patient’s headache improved substantially. The patient...
Bilateral tonsillar herniation into the foramen magnum. Fluid collections (arrows) in the head MRI scan again demonstrating bilateral subdural fluid collections.

The etiology of these lesions remains unclear, although expert opinions have suggested traumatic, inflammatory, hydrostatic, or congenital causes. Unlike meningeal diverticula or Type I meningeal cysts, no clear association with neurocutaneous or connective tissue syndromes has previously been described. One may hypothesize, however, that the same mechanisms responsible for dural ectasia seen in neurofibromatosis and connective tissue disorders (that is, scalloping of the surrounding osseous structures) may also contribute to the bone erosion and cyst expansion seen with giant Tarlov cysts. In our case, the patient's imaging demonstrated multiple other peri-neural cysts as well as profound bone remodeling, suggesting a systemic rather than isolated phenomenon (Fig. 2C and D). The paucity of reported giant perineural cysts in the context of neurocutaneous disorders makes further comments regarding the possibility of a shared pathophysiology difficult.

Clinical presentation of Tarlov cysts may include focal radiculopathy, dyspareunia, coccydynia, and bowel and/or bladder incontinence. Hefti and Landolt suggested that small, focal, true Tarlov cysts not in direct communication with the thecal sac contents are more likely to become symptomatic due to a 1-way valve-like mechanism that entraps CSF within the cyst. This observation is in contrast to meningeal diverticula, where CSF is in free communication and able to reenter the thecal sac, thereby relieving compression on adjacent structures. However, very large Tarlov cysts have also been reported to demonstrate free flow of CSF between the subarachnoid space and cyst body. Tarlov cysts that rupture may also present with intracranial hypotension and are thought to account for most incidences of spontaneous intracranial hypotension. Far from being a benign entity, intracranial hypotension can result in significant neurological deficits, including obtundation and death.

Radiographic features suggestive of this diagnosis include pachymeningeal thickening and enhancement, as well the presence of subdural fluid collections. Brightbill et al. noted the presence of subdural fluid collections only in the presence of pachymeningeal thickening, but not in the converse state, which suggests that the fluid collections represented a more severe state of hypotension. Other radiographic features include descent of the cerebellar tonsils and obliteration of the prepontine cisternal space. Although our patient presented with no focal neurological deficits other than headache, the severity of her symptoms, radiographic findings, and the recognition of the potential morbidity associated with her condition did support prompt intervention once it was apparent that conservative management was unsuccessful.

Fistulization of the spinal subarachnoid space is a known phenomenon, most commonly associated with trauma. Diagnosis is often protracted—neurological changes may be difficult to assess in the context of patients with complex, multiple injuries. The incidence of spinopelvic subarachnoid fistulization is exceedingly rare, with only a single previously reported case. In that case, the patient was also a woman who underwent marsupialization for what was thought to be a pelvic cyst and subsequently developed postural intracranial hypotension. She similarly required operative management of her fistula.

A variety of treatments has been described for intracranial hypotension of differing causes. Therapies such as epidural blood patching, injection of a fibrin sealant, and surgical ligation are mainstays of treatment of idiopathic intracranial hypotension. However, these were believed unlikely to be successful given the sheer size of the cyst and fistula in our patient. Percutaneous therapies were precluded by what was perceived to be difficult access to the cyst and fistulous point. It was concluded that a pedicled omental flap would offer the best chance for successful treatment. The use of pedicled omental flaps is

Discussion

Tarlov or perineural cysts are common but rarely symptomatic lesions. They most often arise in the sacral spine from the S-2 or S-3 dorsal nerve roots. They are classified as Type II meningeal cysts, and large population studies have reported them in 4.6% of the population. The etiology of these lesions remains unclear, although expert opinions have suggested traumatic, inflammatory, hydrostatic, or congenital causes. Unlike meningeal diverticula or Type I meningeal cysts, no clear association with neurocutaneous or connective tissue syndromes has previously been described. One may hypothesize, however, that the same mechanisms responsible for dural ectasia seen in neurofibromatosis and connective tissue disorders (that is, scalloping of the surrounding osseous structures) may also contribute to the bone erosion and cyst expansion seen with giant Tarlov cysts. In our case, the patient’s imaging demonstrated multiple other peri-neural cysts as well as profound bone remodeling, suggesting a systemic rather than isolated phenomenon (Fig. 2C and D). The paucity of reported giant perineural cysts in the context of neurocutaneous disorders makes further comments regarding the possibility of a shared pathophysiology difficult.

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Numerous groups have reported prior successes in treating traumatic subarachnoid fistulas with this and other pedicled tissue flaps.\textsuperscript{2,8,18}

### Conclusions

Although uncommon, large presacral Tarlov cysts should be included in the differential when examining patients with a pelvic lesion. General, urological, and gynecological surgeons should be mindful of the possibility of such cysts and investigate thoroughly prior to surgical intervention. A CT scan will assist in delineating the relationship of a possible intraabdominal Tarlov cyst to the surrounding bony structures and may demonstrate expansion of the adjacent neural foramen. An MRI scan will further delineate the soft-tissue structures surrounding intraabdominal cysts. Flow sequences may demonstrate CSF continuity of large cysts with the thecal sac. If a perineural cyst is ruptured, there exist multiple options for management. Small leaks may be treated nonoperatively with patient positioning, CSF diversion via an external drain, and waiting for the cyst wall to repair itself. Failure of spontaneous recovery or clinical worsening may require further intervention in the form of an epidural blood patch or injection of fibrin glue into the cyst. If the leak

**Fig. 2.** A: Preadmission CT scan of the abdomen demonstrating a large, left-sided presacral perineural cyst arising from the neural foramen. Of note, there is also expansion of the right neural foramen and bony canal. As well, these findings suggest a right perineural cyst that is within the neural foramen. B: Admission CT scan of the abdomen at approximately the same level as in panel A, demonstrating a shrunken left perineural cyst (arrow). The right perineural cyst remains constant in size. C: Abdominal MRI scan, T2 sequence, at approximately the same level as in panels A and B, demonstrating the marsupialized left perineural cyst (upper arrow), with blood products within the lesion (lower arrow). The right perineural cyst can also be better appreciated, but it has not changed significantly in size in comparison with pre-presentation imaging. D: Sagittal CT scan of the abdomen demonstrating the marsupialized perineural cyst (arrow).
IS large or otherwise inaccessible percutaneously, operative repair may be required. There are a variety of surgical approaches and techniques, including primary repair and free and pedicled tissue flap transfers. The selection of the most appropriate technique depends on a number of factors including the severity of the leak, the patient’s anatomy, and the surgeon’s comfort with each approach. Neurosurgical consultation and/or involvement is recommended prior to definitive treatment.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Sahjpaul, Tu. Acquisition of data: Tu. Analysis and interpretation of data: Tu. Drafting the article: Sahjpaul, Tu. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Sahjpaul. Administrative/technical/material support: all authors. Study supervision: Sahjpaul.

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