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CASE REPORT

Giant cystic sacral schwannoma mimicking tarlov cyst: a case report

Mark A. Attiah[1] · Peter P. Syre[1] · John Pierce[1] · Elizaveta Belyaeva[2] · William C. Welch[1]

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Abstract
Purpose To present a rare case of a giant schwannoma of the sacrum mimicking a Tarlov cyst.
Methods A 58-year-old woman had a 1-year history of low back pain. MRI revealed a large cystic mass in the sacral canal with bony erosion. Radiological diagnosis of Tarlov cyst was made.
Results The patient underwent surgical treatment for the lesion, which revealed a solid mass. Histopathological examination of the tumor confirmed the diagnosis of schwannoma. The postoperative course was uneventful and the patient has had significant improvement in her pain 1 month postoperatively.
Conclusion Giant cystic schwannoma of the sacrum is a very rare diagnosis overlooked by practitioners for more common cystic etiologies, but its treatment is significantly different. Care should be taken to include this diagnosis in a differential for a cystic sacral mass.

Keywords Giant Schwannoma · Sacral · Tarlov cyst

Introduction

Schwannomas are benign, slow-growing neoplasms that arise from myelinated nerve sheaths. Spinal schwannomas are the most common intradural extramedullary tumors, accounting for 25 % of all primary spinal tumors [1, 2]. However, sacral schwannomas are very rare, accounting for only 1-5 % [3]. Because of their indolent growth, the mobility of the sacral nerve roots and the generous width of the sacral canal, these tumors can reach large sizes before becoming symptomatic. Most schwannomas are solid or heterogenous tumors, occasionally exhibiting hemorrhagic, calcific, or fibrotic degenerative changes. Cystic changes, however, are exceedingly rare, with only a few cases being reported in the literature [4–6]. We present a 58-year-old woman with cystic giant sacral schwannoma, mimicking an atypical Tarlov cyst.

Report of the case

A 58-year-old woman presented with a 1-year history of left buttock and lower back pain. Occasionally she experienced left leg pain and rarely tingling sensations down into her toes in a general sciatic distribution. Her symptoms were aggravated by sitting and were progressively worse throughout the day. Her buttock pain was improved with laying supine. General physical examination revealed some loss of lumbar lordosis. Neurologic examination revealed bilateral patellar and Achilles hyperreflexia. MRI of the lumbar spine without contrast revealed an expansile 2.2 × 3.5 × 3.3 cm cystic lesion expanding the sacrum beginning at S2-3 (Fig. 1). The mass involved the S3 segment on the left side principally, though the mass crossed the midline and also involved the right-sided S3 nerve root as well. The signal showed a slightly different consistency than CSF around the nerve roots. Computed tomography (CT) scan with contrast revealed a well-circumscribed, hypodense, and non-enhancing cystic structure occupying the entire paramidline sacral central canal extending from the superior aspect of S2 to the inferior

✉ Mark A. Attiah
mattiah12@gmail.com

[1] Department of Neurosurgery, University of Pennsylvania, 3400 Spruce St., Philadelphia, PA 19104, USA

[2] Department of Pathology, University of Pennsylvania, Philadelphia, USA

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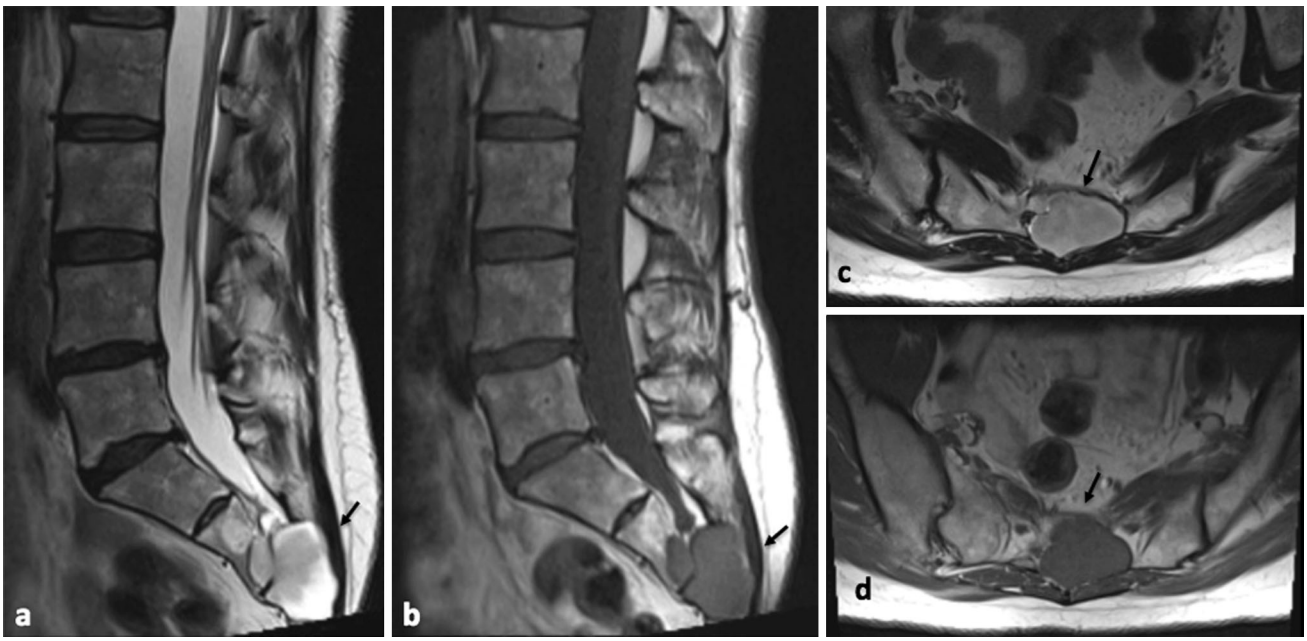

Fig. 1 MR images demonstrating large cystic lesion in the sacrum (*arrow*). **a** Sagittal T2-weighted image. **b** Sagittal T1-weighted image. **c** Axial T2-weighted image. **d** Axial T1-weighted image

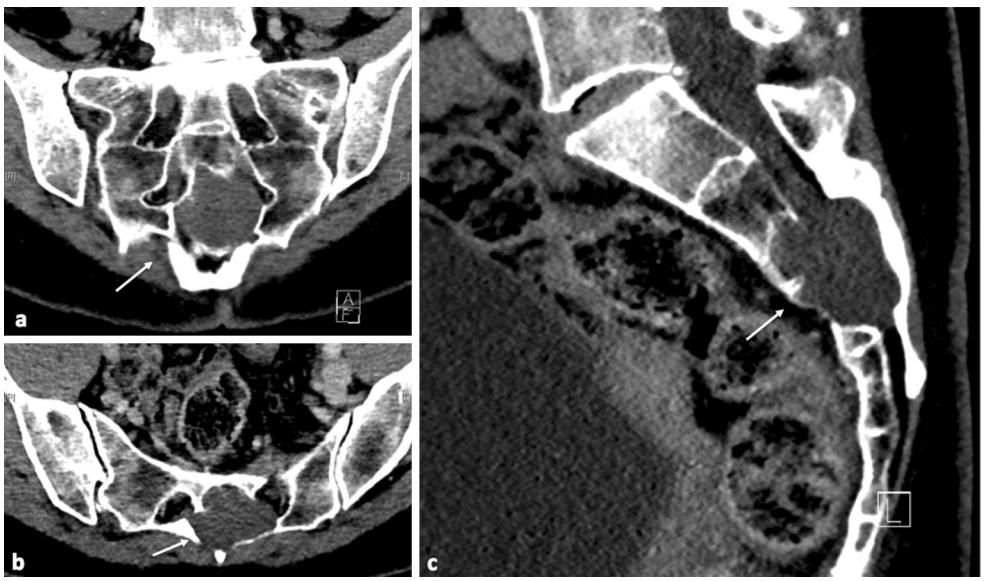

Fig. 2 CT scan with contrast demonstrating a large homogenous non-enhancing soft tissue mass (*arrows*) with sclerotic margins. **a** Coronal. **b** Axial. **c** Sagittal

aspect of S3 causing mild expansion and scalloping of S2-S3 and posterior column (Fig. 2).

The preoperative diagnosis was a Tarlov cyst. The patient underwent left sacral laminectomies of S1, S2, and S3 with mass resection and reconstruction with paraspinal muscle flap. After removing the thinned sacral bony lamina, a solid mass could be identified underneath a thinned dural layer. The mass was rubbery, with the consistency of fibrin glue, and associated with the left S2 nerve root within and external to the sacral dura (Fig. 3). The dura was slightly torn from bony removal but no obvious fluid leaks could be identified. Normal roots traversing the mass could be seen both superiorly and inferiorly and were stimulated using neuromonitoring. These roots were the sacral roots of S3 and S1 and were not involved in the mass. However, despite the mass's minimal attachment to

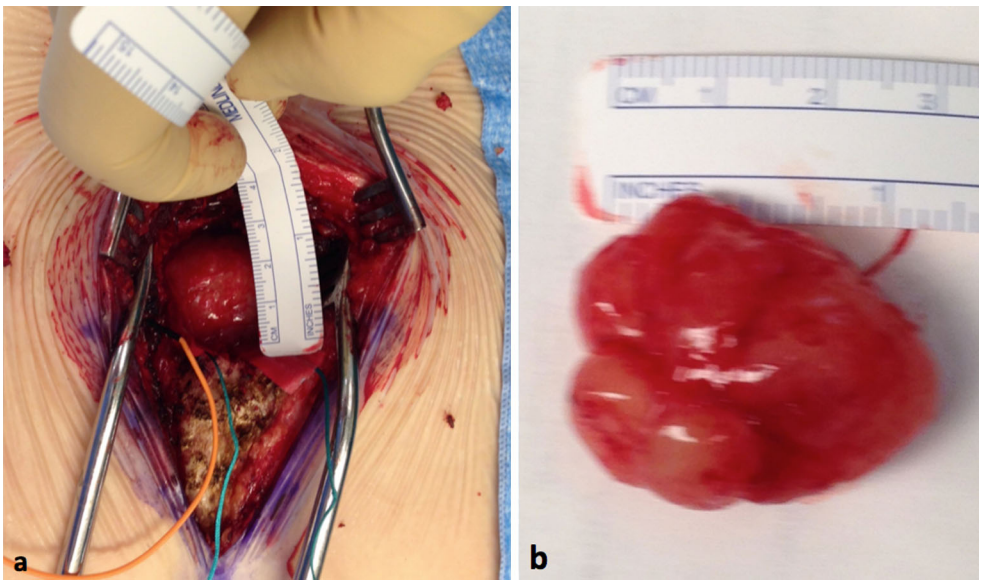

Fig. 3 **a** Intraoperative view of giant sacral cystic schwannoma. **b** Pathologic specimen of tumor

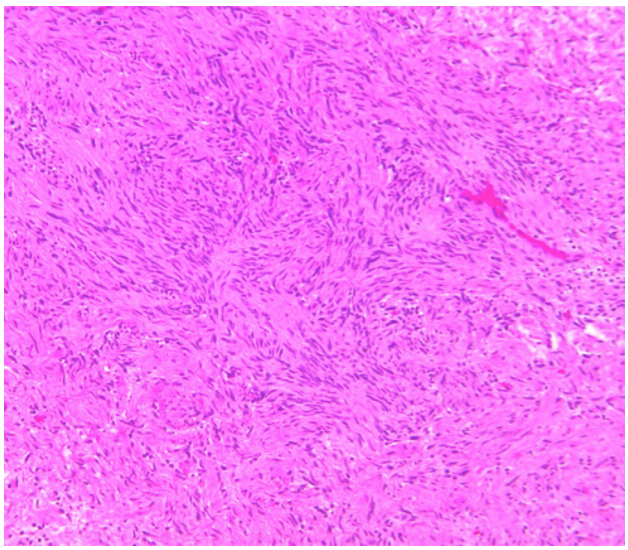

Fig. 4 Histopathology of tumor, demonstrating hypercellular Antoni B area tissue with nuclear palisading in a neurofibromatous background

the S2 nerve, the nerve itself was atrophic and nonfunctional as demonstrated by direct current stimulation during intraoperative monitoring. This was dissected off the dorsal portion of the mass but had areas where no adequate plane was identified between the mass and these root-like structures. The mass and some of the associated nerve was removed, along with a smaller lesion with a similar appearance. Closure was performed with paraspinous muscle flap procedure.

Histopathological examination (Fig. 4) showed Antoni B area tissue with nuclear palisading in a neurofibromatous background, supporting the diagnosis of cystic schwannoma.

The patient tolerated the procedure well. The postoperative course was uneventful and was discharged home 3 days after surgery without any complications. At 1-month follow-up, the patient reported that preoperative left buttocks pain frequency decreased from constant to intermittent and from focal to more diffuse. She still had mild residual left leg pain, but she was able to walk several miles not limited by pain.

Discussion

Nerve sheath tumors such as schwannomas and neurofibromas account for about 30 % of all intraspinal tumors [7], usually arising in the lumbar region with a preference for sensory nerves and the dorsal roots [8]. Sacral schwannomas are very rare entities, with only about 50 cases reported in the literature worldwide [3]. Because of their slow-growing nature, and the mobility of the nerve roots in the sacral canal, symptoms do not manifest until the tumor grows to be quite large [9]. Presenting symptoms are most commonly progressive lower back pain, numbness or paresthesias, neurological deficits, and failure of conservative therapy such as physical rehabilitation [10]. However, acute lower back pain may also be caused nerve root torsion or hemorrhage within the tumor itself [11, 12]. Nonspecific abdominal pain has also been reported [13].

Sacral schwannomas with a predominantly cystic component are rarer still. Postulations as to the etiology of this cystic component have been put forth previously, including

ischemic necrosis, hemorrhage within the tumor, mucinous degeneration, and the formation of micro cysts. The differential for a large cyst in this location is extensive, including abscess, arachnoid cyst, ependymoma, epidermoid cyst, cystic lymphangioma, cystic teratoma, and cystic meningioma. MRI is the most useful imaging modality for diagnosis of these tumors. Schwannomas have a low-to intermediate signal intensity on T1-weighted images and a heterogenous appearance with focal areas of hyper- and hypo-intensity corresponding to cyst formation on T2-weighted images [14, 15]. With contrast, rim enhancement of an intradural extramedullary mass is characteristic of a schwannoma [15]. CT is also helpful and can show a nonspecific, well-circumscribed lesion with a low or heterogenous signal, sometimes with central necrosis [16]. When considering the differential for a cystic mass in this region, however, there are no distinguishing radiographic features that are pathognomonic for cystic schwannomas. Tarlov cysts are also primarily located in the S1–S4 region, form within the nerve root sheath at the dorsal root ganglion, and are filled with fluid that is radiographically similar to CSF [17]. These characteristics, plus the patient's symptoms of low back and buttock pain lead to a preoperative diagnosis of Tarlov cyst by multiple radiologists and neurosurgeons. Because the treatment approaches for Tarlov cysts are radically different (i.e., cyst fenestration vs. complete resection vs. no treatment), cystic schwannomas should be considered in the differential of a cystic osteolytic mass in the sacral canal.

Histopathologic examination is necessary for proper diagnosis as Tarlov cysts that have been previously treated with fibrin glue injections can closely resemble solid tumors. The classic histologic findings of Schwannomas are spindle-shaped cells with pale, eosinophilic cytoplasm arranged in Antoni A (compact, hypercellular, and well-organized spindle cells in a palisading formation) or Antoni B (hypocellular, pleomorphic cells with a predominantly myxoid cytoplasm) patterns [18]. Verocay bodies, also characteristic of schwannomas, can aid in definitive diagnosis [18].

Unlike Tarlov cysts, the optimal treatment for giant spinal schwannoma is complete excision. This lessens of the possibility of recurrence seen with subtotal resections [6, 19, 20]. Though not a common occurrence, recurrent tumors necessitate a second operation with the associated risks. Navigating scar tissue and the loss of anatomical landmarks increase the likelihood of nerve root damage and neurological deficits. The nerve root associated with the tumor was sacrificed in this case, however, complete excision without damage to an associated nerve root is indeed possible as entrapment only occurs in 50 % of cases [21–23].

The surgical approach is highly dependent on the extent of tumor involvement. We used a posterior approach, as this was suspected to be the more common entity, Tarlov cyst. A posterior approach can be used in sacral schwannoma if the tumor does not extend beyond the anterior vertebral body. However, in sacral schwannomas that do extend anterior to the vertebral body a combined approach is warranted. There are several characteristics to consider when approaching a large tumor in this area. The tumor capsule may be fragile or adherent to surrounding structures, and the arachnoid planes may be difficult to identify. Further, the increased vascular supply to the sacral region may present a large amount of intraoperative bleeding. The bony erosion caused by such a large tumor can render the spinal column unstable after excision, necessitating spinal stabilization. Various stabilization techniques have been employed after treatment of spinal schwannomas [6, 24]. In our patient, complete excision was successfully performed without the use of any spinal reconstruction or stabilization procedure as the anterior sacral bone remained intact.

The diagnosis of a sacral cystic schwannoma is exceedingly rare, and can mimic other entities on presentation and radiographic imaging. Care should be taken to include this type of tumor in the differential diagnosis of a cystic sacral lesion, as the treatment for a schwannoma can be significantly different from another such as a Tarlov cyst.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest

Informed consent The patients and/or their families were informed that data from the case would be submitted for publication, and gave their consent.

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