Giant cystic intradural schwannoma in the lumbosacral region: a case report

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ABSTRACT
We report a case of a giant cystic intradural schwannoma of the lumbosacral region in a 30-year-old man who presented with a 2-year history of non-specific lower back pain. Lateral radiographs demonstrated scalloping of the posterior wall of L5 and the upper sacrum. Magnetic resonance imaging revealed a 12x2.3-cm intradural multi-septated cystic lesion extending from L3 to S2 with predominant hypointense signal on T1-weighted images and a mixed signal on T2-weighted images. There was heterogeneous rim enhancement of the retrosacral portion of lesions following the administration of gadolinium contrast. The tumour was completely excised. Histological investigation confirmed the diagnosis of cystic schwannoma with alternating hypercellular (Antoni A) and hypocellular (Antoni B) areas in a fibrillar background. The patient had complete relief of symptoms and remained asymptomatic after 2 years of follow-up.

Key words: lumbosacral region; nerve sheath neoplasms; neurilemmoma; spinal cord neoplasms

INTRODUCTION
Schwannoma (neurilemmoma) is a benign slow-growing encapsulated tumour arising from the myelinated nerve sheaths. Schwannomas, together with meningiomas, are the most common intradural tumours. Degenerative changes like haemorrhage, calcification, and fibrosis are commonly seen in schwannomas, but cystic changes are rare. Such tumours have been reported in the orbital region, olfactory groove, tentorial hiatus, posterior cavernous sinus, presacral region, pancreas, maxillary sinus, spinal cord, lesser sac, and within the ventricular system. Only one case of giant cystic schwannoma in the lumbosacral region has been reported. We report another such case in a 30-year-old man.
CASE REPORT

In March 2005, a 30-year-old man presented with a 2-year history of progressive lower back pain radiating to both limbs. The pain in the lower back was aggravated by lying supine and walking, while relieved by sitting. He had no history of antecedent trauma or constitutional symptoms. Physical examination revealed lumbar paravertebral muscle spasm, decreased spine movements, and negative root tension signs. Neurological tests revealed the absence of bilateral ankle reflexes. No cutaneous stigmata of neurofibromatosis were seen. Lateral radiographs demonstrated scalloping of the posterior wall of L5 and the upper sacrum (Fig. 1). Magnetic resonance imaging (MRI) revealed a 12x2.3-cm cystic lesion filling almost the entire spinal canal extending from L3 to S2 with septation, scalloping of the adjacent vertebral bodies, and a hypointense signal on T1-weighted images, but mixed signal on T2-weighted images. There was heterogeneous rim enhancement of the retrosacral portion of the cystic lesion following the administration of gadolinium contrast (Figs. 2 and 3).

Figure 1  A lateral radiograph of the lumbosacral spine showing scalloping of the posterior wall of L5 and upper sacrum with no apparent widening of the intervertebral foramen.

Figure 2  (a) Axial and (b) coronal T2-weighted images of L5 demonstrating multiloculated cystic lesions occupying almost the entire canal (arrows).

Figure 3  Mid-sagittal magnetic resonance images reveal intradural tumour occupying the spinal canal from L3 to S2 showing (a) hyperintensity on T2-weighted imaging, (b) hypointensity on T1-weighted imaging and (c) rim enhancement in the retrosacral portion on T1-weighted imaging.
Bilateral laminectomy (L3 to S2) was performed using a posterior midline approach to expose the dura, which was found to be thinned out, tense, and without pulsation. Under microscopic visualisation, a midline durotomy was performed and a well-defined, encapsulated, yellowish, soft, friable, cystic tumour was found loosely adherent to the arachnoid, displacing the roots peripherally. The tumour was easily separated from the dura. The tumour was dissected intact by cutting the vascular pedicle at S1 with bipolar cautery.

Histopathological examination showed alternating Antoni A (hypercellular) and Antoni B (hypocellular) areas in a fibrillar background without any axons suggestive of cystic schwannoma (H&E, x100).

Various names, including perineural fibroblastoma, neurilemmoma, neurinoma, neurilemoma, and peripheral glioma, have been used. In the spinal region, schwannomas have a predilection for sensory nerves and tend to arise from the dorsal roots.13 Nerve sheath tumours, such as schwannomas and neurofibromas, account for 30% of all intraspinal masses.14 Intraspinal schwannomas are most frequently seen in the lumbar region with a predilection for the lower cervical and thoracolumbar junction.13 Spinal schwannomas may be well-circumscribed, intradural or extradural, or combined intra-extradural. The percentage of purely intradural nerve sheath tumours increases from 8% in the upper cervical to 80% in the thoracolumbar region.15 This may be explained by the anatomic features of the spinal nerve roots, which have a longer intradural component at the caudal portion of the spinal axis. Multiple lesions are common in neurofibromatosis or schwannomatosis,16 which is characterised by the presence of multiple schwannomas without the stigmata of neurofibromatosis. Coexistence with other tumours especially meningioma can be seen in patients with neurofibromatosis.17 Lumbago is the usual symptom. Features of cauda equina tumours include pain on lying in a recumbent position, progressive pain and neurological deficits, involvement of nerve roots, and failure of conservative therapy.18 These features help differentiate tumours from mechanical causes of back pain. Delayed presentation is common because these tumours are slow growing, the surrounding anatomic environment is permissive, the presenting symptoms are non-specific, and the affected patients’ are young and otherwise healthy. Intradural schwannomas may, in rare cases, lead to intracranial subarachnoid haemorrhage19 or hydrocephalus.20 They may present as acute lower back pain secondary to nerve root torsion21 or traumatic intratumoural haemorrhage.22 Even more rarely, they may present as non-specific abdominal pain.23 Tumours arising in the cauda equina or around the conus medullaris can become larger than other spinal tumours because of the relative mobility of the roots and the wide diameter of the spinal canal.24 No radiographic findings are pathognomonic for intraspinal schwannomas. However, imaging studies may reveal widening of the neural foramen, erosion of the pedicle, increased interpedicular distance, and scalloping of the adjacent vertebral body. MRI is the preferred imaging modality. Schwannomas generally have low-to-intermediate signal intensity on T1-weighted images. On T2-weighted images, they may be heterogeneous with focal areas of hyperintensity.

DISCUSSION

Schwannomas are tumours arising from the embryonic neural crest cells of the nerve sheaths of peripheral and cranial nerves. The first case was reported in 1910.12 Figure 4 Photomicrograph of the specimen showing alternating Antoni A (hypercellular) and Antoni B (hypocellular) areas in a fibrillar background without any axons suggestive of cystic schwannoma (H&E, x100).
and hypointensity, corresponding to cyst formation, haemorrhage, dense cellularity, and collagen deposition. Very high intensity regions seen on T2-weighted images correspond to cystic degeneration with surrounding collagenous fibrous tissue. Schwannomas do not always have the typical T2 appearance of benign neurofibromas, specifically a hyperintense rim of myxomatous tissue surrounding a hypointense centre of fibrocollagenous tissue. With gadolinium contrast, the tumour is dense and heterogeneous, though smaller lesions may be homogeneous. In particular, rim enhancement of an intradural extramedullary tumour suggests the diagnosis of schwannoma.

Definitive diagnosis is based on histological analysis. Schwannoma in its classic form consists of spindle-shaped cells with pale, eosinophilic cytoplasm arranged in 2 characteristic patterns: Antoni A (compact, hypercellular, well-organised spindle cells in a palisading pattern) and Antoni B (hypocellular, loose-textured pleomorphic cells with predominantly myxoid cytoplasm). The hallmark histological feature of the schwannoma is the Verocay bodies. Immunohistochemical staining can further aid in the diagnosis as benign schwannomas show diffuse immunoreactivity for S-100 protein.

Histological variants have been described. Ancient schwannoma, a rare variant first described in 1951, is characterised by degenerative changes of cyst formation, calcification, haemorrhage, and fibrosis with cytological atypia. They are typically asymptomatic and difficult to diagnose. These degenerations are usually seen in part of the tumours. The presence of a predominantly cystic schwannoma in the lumbosacral region is rare. These cystic changes are likely attributable to mucinous degeneration, ischaemic necrosis, haemorrhage, and the formation of microcysts. Cellular schwannomas differ from the classic type in that the dense Antoni A pattern comprises ≥90% of the tumour area with a more uniform pattern, a lack of Verocay bodies, and frequent lymphocytic infiltration. Melanotic schwannoma is another rare variant comprising melanin-producing cells with ultrastructural features of Schwann cells.

Complete excision of tumour may prevent recurrence. Although schwannomas originate from nerve tissue, only 50% of cases have a direct relation with a nerve. Hence complete excision without sacrificing nerve roots is feasible in most of the contained non-invasive varieties. Increased vascularity in the sacral region can be tackled easily with intra-operative haemostasis when anticipated. Surgical treatment of cystic schwannomas can be very demanding because of the adhesion of the tumour capsule to the surrounding structures, fragile tumour capsules, and difficulty in identifying the arachnoidal planes. Early identification of the arachnoidal planes without opening of the cyst and sharp dissection may be useful. Complete excision without resultant neurological deficits may be feasible provided that there is no entrapment of nerve roots.

Prognosis is usually excellent with the exception of the melanotic variant, malignant forms, and cases of neurofibromatosis.

The differential diagnosis of such a large cyst includes a cystic schwannoma, cystic neurofibroma, ependymoma, epidermoid cyst, arachnoid cyst, cystic lymphangiomas, cystic teratoma, and cystic meningioma. Histopathological examination remains the mainstay of differentiation as clinico-radiographic features can be indistinguishable.

10 cases of predominantly cystic intradural schwannoma have been reported. Only one described the giant configuration. It was incompletely excised and necessitated additional anterior reconstruction and posterior stabilisation for the grossly eroded and osteoporotic adjacent vertebrae. In our patient, complete excision was successful. Intraspinal tumours of such magnitude usually invade surrounding bone or present with neurological deficits. A successful surgical outcome depends on early diagnosis (before neurological damage has occurred), meticulous investigation (on any intractable non-specific lower back pain), and complete excision.

REFERENCES