Fluid-fluid levels in cystic lumbosacral schwannomas: a report of three cases

ABSTRACT
Magnetic resonance imaging features of three benign cystic lumbosacral schwannomas, which showed multiple fluid-fluid levels, are described. One of the tumours showed bone destruction with fluid-fluid levels that closely mimicked an aneurysmal bone cyst. Pathological examination confirmed haemorrhage as the cause of fluid-fluid levels in the tumours. Though a rare finding, fluid-fluid levels with bone destruction can also be caused by nerve sheath tumours and hence must be included in the list of differential diagnoses of spinal tumours.

Keywords: cystic schwannoma, fluid-fluid levels, haemorrhagic tumour, lumbosacral schwannoma, schwannoma

INTRODUCTION
Schwannomas represent 8% of primary intracranial tumours and 29% of primary intraspinal tumours.1 These tumours are benign, usually well-defined, arise from Schwann cells and are described in virtually all body locations. The lumbar region is one of the most common sites for the occurrence of spinal schwannomas. Conti et al reported a frequency of 48% of schwannomas in the lumbar region.2 Schwannomas can often be large and go unnoticed for years, mainly due to their indolent growth, until a compressive effect is produced upon the spinal cord or nerve roots. Schwannomas can show a variety of degenerative changes, such as cyst formation, calcification, haemorrhage and hyalinisation.3

We report three cases of large cystic schwannomas producing extensive destruction and expansion of the vertebra with multiple fluid-fluid levels. Because of the multiple fluid-fluid levels and bone expansion, one of these cases had the appearance of a primary bone tumour like an aneurysmal bone cyst. These cases illustrate that benign spinal schwannomas, due to cystic degeneration and repeated haemorrhage, can present as cystic masses with bone destruction and fluid levels and hence, should also be considered in the differential diagnosis.

CASE REPORTS
Case 1
A 40-year-old woman presented in March 2002 with a dull aching low backache, occasionally radiating to the bilateral thigh, for six years. She developed slow progressive difficulty in climbing stairs and getting up from the sitting position over the past four years. She had numbness of both lower limbs, which gradually ascended to the level of the groin. Bowel and bladder habits were normal. Right lower limb and proximal left hip flexors power were 4/5. Bilateral extensor hallucis were 4/5 and knee and ankle jerk on the right side were absent. She was noted to have decreased pain and touch sensation in the L3 dermatome.

Radiographs of the lumbar spine showed erosion of the L3 pedicle and widened interpedicular distance. Magnetic resonance (MR) imaging of the lumbar spine showed a heterogeneous solid and cystic mass causing cord compression and extending through the L3 and L4 neural foramen. A diagnosis of lumbar schwannoma was made, and subsequent surgical excision and histopathological examination confirmed the diagnosis of schwannoma. The patient was symptom-free for the next three years, after which she developed a radiating pain and paresthesias in both the lower limbs. Examination revealed power of 4/5 in both lower limbs, absent knee and ankle reflexes in both the lower limbs with bilateral plantar flexor response. Tandem walking was impaired and she had impaired sensation in the L4, L5, S1 and S2 dermatomes, predominantly affecting the right side.

Contrast-enhanced MR imaging of the lumbar spine showed a well-defined lobulated cystic lesion appearing hyperintense on T2- and hypointense on T1-weighted images, causing eccentric erosion of the L4 vertebral body and right pedicle, and extending into the paravertebral region displacing the psoas muscle laterally. The cystic mass had an intraspinal component, which was seen displacing the cauda equina and compressing it. The cystic lesion had multiple fluid-fluid levels with the dependant part showing hypointensity, suggesting the possibility of haemorrhage. There was thin mild enhancement of the septations and the cyst wall (Figs. 1a–d). Computed tomography (CT) showed destruction of the L4 vertebral body and right pedicle (Fig. 1e). With these imaging findings, the possibility of a primary bone tumour like an aneurysmal bone cyst was considered. However, since there was a previous history of excision for neurogenic tumour in the same region, the possibility of recurrence of the tumour was also considered. She was re-explored and was found to have recurrent schwannoma that was excised and confirmed on histopathology (Fig. 1f).
Case 2
A 25-year-old woman presented with low backache and radiating pain in both lower limbs for nine months which was aggravated by walking, coughing and sneezing. She noticed a slowly increasing swelling in the lower back, one month after the onset of the pain. She did not have any motor, sensory or bladder and bowel symptoms. Examination revealed left hip abduction of power 4/5 and sluggish bilateral knee jerks. Radiographs of the lumbosacral spine showed smooth enlargement of the sacral canal with scalloped margins. MR imaging of the lumbosacral spine showed a large multiloculated cystic lesion in the dorsal sacral region at the level of L5–S1, producing scalloping of the posterior margin of the vertebral body. The mass was of mixed intensity on T1- and hyperintense on T2-weighted images. Multiple fluid-fluid levels were seen in the dependant part, with the lower layer being hypointense, suggesting haemorrhage. Post-contrast MR images showed thin peripheral enhancement (Figs. 2a–d). The lesion was diagnosed as a cystic schwannoma with haemorrhagic changes, which was confirmed at surgery. Histopathology of the excised mass was benign schwannoma (Fig. 2e).

Case 3
A 65-year-old woman presented with gradually progressive weakness of the right lower limb, followed by the left lower limb over the past year. The weakness involved both proximal and distal muscles, which subsequently made her bed-bound and she had to be catheterised for urinary retention. She did not have any bowel symptoms or sensory impairment. On examination, her reflexes were absent in both lower limbs, power was 4/5 in the hip flexors and extensors, 2/5 in adductor and abductor muscles of the hip, 4/5 in knee flexors and extensors, 1/5 in dorsiflexors and 3/5 in plantar flexors of the ankle joint on both sides. Vibration senses were impaired on both sides; the rest of the sensory examination was normal. Anal tone was poor.
Radiographs showed extensive posterior scalloping of the lumbar vertebra with an expanded spinal canal, destroyed pedicles and enlarged neural foramen at multiple levels. MR imaging of the lumbar spine revealed an expansile, multiloculated cystic lesion extending over several lumbar vertebrae, causing extensive scalloping of the posterior vertebral body, with destruction of pedicles, lamina and extending through the neural foramen into the posterior soft tissue. Two cysts in the superior and inferior aspect were hyperintense on T1- and T2-weighted sequences, probably representing high proteinaceous or haemorrhagic fluid. The cysts were variable in size and a few showed fluid-fluid levels with hypointense layering, possibly due to haemorrhage. There was enhancement of the septae and cyst wall (Figs. 3a–d). A diagnosis of cystic schwannoma was made which was confirmed on surgery and histopathology (Fig. 3e).

**DISCUSSION**

Schwannomas are benign, slow-growing, well-encapsulated tumours arising from Schwann cells but do not incorporate nerve roots. Most commonly, these tumours arise around the peripheral nerve roots in the extradural space. Women and men are equally affected, and are usually seen in the fourth to sixth decades of life. There is a 5% association with neurofibromatosis (NF)-1. Ancient schwannoma is a rare variant which was first described by Ackerman and Taylor in 1951, and predominantly consists of Antoni B tissue. This is characterised by degenerative changes such as cyst formation, calcification, haemorrhage, fibrosis and cytological atypia. It is typically asymptomatic and difficult to diagnose in the absence of clinical symptoms. Symptoms are usually due to its pressure effects on nerve roots or adjacent organs.

Histologically, two types of tissues are seen in schwannomas, viz. Antoni A and Antoni B, that represent areas of myxoid matrix. These tumours often show striking cytological atypia but mitotic figures are rare. Waxing and waning of the tumour size is attributed to fluctuations in its cystic content. Incompletely
excised lesions show recurrence over a period of time. High recurrence rates are observed in cases of intracranial, intraspinal and sacral schwannomas, plexiform neurofibromas and giant schwannomas. In rare situations, these tumours can be locally aggressive and show cellular atypia, mitosis, increased cellularity and bony extension.\(^5\)

Cystic degenerations occurring in schwannomas are rarely reported in schwannomas of the orbital region, olfactory groove, tentorial hiatus, posterior cavernous sinus, presacral region, maxillary region, intraspinal and intraventricular locations. They are also observed in schwannomas arising from the vestibular or vagal nerve, jugular foramen and thoracic neurilemmomas.\(^6\)\(^-\)\(^8\) Five cases of cystic nerve sheath tumours in the lumbar region and its MR imaging characteristics have also been described.\(^14\) Borges et al described a case of intradural cystic lumbosacral schwannoma and suggested that cystic changes are rare in lumbosacral schwannomas.\(^15\)

Fluid-fluid levels are seen in many bony as well as soft tissue tumours, and can be a non-specific finding. Layering in cystic tumours can be due to repeated haemorrhage into the tumour, uncommonly due to fat, calcium or protein-rich fluid.\(^16\)\(^-\)\(^18\) Tumour necrosis, thrombosis of the vessels with resultant necrosis plus haemorrhage or tumour neovascularity have been described as possible mechanisms of haemorrhage in the schwannoma.\(^19\)\(^-\)\(^20\) Blood in these cases often does not clot and forms a layer depending on the viscosity and proteinaceous content of the cyst fluid.\(^21\) This often results in the formation of fluid-fluid levels. The Antoni B portion of the schwannoma can undergo degeneration to form cystic areas that can progress to become large cysts.\(^22\) Shiono et al reported two cases of lumbar cystic schwannoma, one of which had fluid-fluid levels. The cyst fluid contained red blood cells, confirming repeated episodes of haemorrhage, leading to necrosis and the formation of fluid levels.\(^23\) Vilanova et al described one case of malignant schwannoma of the lumbar spine with haemorrhage and fluid-fluid levels.\(^24\)

Fluid-fluid levels have been described in acoustic and trigeminal schwannomas, but are very uncommon...
in lumbar schwannomas. In our cases, fluid-fluid levels were seen in benign schwannomas. None of the presented cases had features suggestive of a malignant transformation. The most likely cause of the fluid-fluid levels in our cases was necrosis with repeated haemorrhage into the tumour. The presence of haemorrhagic fluid within the tumour was confirmed during surgery in these cases. We did not find any correlation between the type of Antoni cells and degenerative changes seen within the tumour, as was reported earlier. Also, these tumours were highly vascular and bled profusely during surgery, which might be the reason for the presence of haemorhagic fluid levels on MR imaging. Histopathological studies have shown the presence of multiple prominent vasculatures within these tumours.

Schwannomas causing extensive destruction of the vertebral body are reported to be rare. Our search of the literature found that only four cases of schwannomas causing destruction of the vertebral body have been reported to date. Singrakhia et al described two cases of schwannoma associated with expansile osteolytic destruction of the cervical vertebra. The mechanisms described include secondary erosion of the vertebra, a tumour arising within the bone (intraosseous schwannoma) or a tumour arising in the nutrient canal and growing in a dumbbell configuration. The tumour can grow along the branch of the spinal nerve into the vertebral body through the nutrient foramen and cause destruction of the vertebra. In our first case, the extensive destruction of the vertebra might be secondary to erosion of the vertebral body by the tumour or growth of the tumour along the spinal nerve branch as described by Inoue et al.

Radiological differential diagnoses include primary bone tumours like aneurysmal bone cysts, giant cell tumours (GCT), osteoblastoma, metastases and spinal cord tumours like ependymoma. Aneurysmal bone cysts are expansive lytic tumours that commonly involve the posterior elements but can extend to the vertebral body. These tumours frequently show cystic areas with fluid-fluid levels due to haemorrhage. Aggressive osteoblastomas can appear as expansive lytic lesions with destruction of adjacent bone and soft tissue extension, but the presence of matrix calcifications may help in differentiating them from other similar lesions. Although GCT commonly affects vertebral bodies, they can show extension into posterior elements. GCTs can exhibit fluid-fluid levels, and MR imaging may show low signal intensity due to haemorrhage or increased fibrous content. Rarely, myxopapillary ependymomas can show massive destruction of vertebral bodies with expansion of neural foramina and soft tissue

REFERENCES