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Cystic spinal schwannomas: A short series of six cases. Can we predict them preoperatively?

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Abstract

Background: Spinal schwannomas are benign tumors arising from the spinal nerve root sheaths and are the commonest intradural extramedullary spinal tumors. Though cystic changes in schwannomas are well described, predominantly cystic schwannomas are uncommon lesions and form a different spectrum of conditions as compared with the commonly seen intradural extramedullary solid lesions.

Case Description: We present a case series of six patients with spinal intradural extramedullary cystic schwannomas. Two patients had uniloculated cystic schwannomas, two patients had multi-loculated cystic lesions with thick walls and intralesional septations, and two patients had giant cystic schwannomas, one of which had an extradural extension. We report two cases in which preoperative radiological dilemma was encountered and discuss the differential diagnoses of this uncommon entity.

Conclusion: Cystic spinal schwannomas may be confused with other cystic lesions in the spine, differentiating them preoperatively is important and in this regard, contrast-enhanced magnetic resonance imaging plays a vital role. Frozen section histopathology should be used to identify them at surgery. It is important to detect these lesions at surgery, as total excision is possible and almost always results in good long-term neurological outcome.

Key Words: Benign spinal tumors, cystic schwannomas, spinal schwannomas

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INTRODUCTION

Spinal schwannomas are benign tumors arising from the spinal nerve root sheaths and are the commonest intradural extramedullary spinal tumors.[2] Schwannomas are mostly solid or heterogeneously solid tumors. Though cystic changes in solid schwannomas are well described, predominantly cystic schwannomas are uncommon.[5,6] Differentiating these cystic lesions from similar cystic lesions in the intradural extramedullary space is important, and magnetic resonance imaging (MRI) plays a vital role in this context.

MATERIALS AND METHODS

We present a case series of six patients with spinal intradural extramedullary cystic schwannomas. Two patients had uniloculated cystic schwannomas, two patients had multi-loculated cystic lesions with thick walls and intralesional septations, and two patients had giant

cystic schwannomas, one of which had an extra-dural extension. The clinical and radiological presentations along with the intra-operative findings and post-operative courses of the six patients in this series are represented in Table 1.

Illustrative cases

Case 2

A 50-year-old female presented with complaints of backache, numbness of the right lower limb, and difficulty in walking. On examination, she had motor weakness in both lower limbs (grade 3/5 at the right knee and ankle, and grade 4/5 at the left knee and ankle), and had decreased sensations below the level of L2 dermatome. MRI of the lumbo-sacral spine revealed an intradural cystic space occupying lesion at the level of L2-L4 vertebrae, The cyst exhibited intensity similar to cerebrospinal fluid (CSF) on T1-weighted image (T1WI) and T2-weighted image (t2WI) [Figure 2a, b, d, and e], with rim-enhancement on contrast administration [Figure 2c]. The patient underwent L2-L4 laminectomy and total excision of the lesion. Intraoperatively, on opening the dura, a cystic lesion with translucent walls was seen displacing the roots on either side [Figure 2f]. Cyst was decompressed and lesion was excised maintaining the arachnoid plane [Figure 2g and h]. The patient had an uneventful recovery and motor functions and sensations improved to normal at 6 months follow-up.

Case 6

A 25-year-old female presented with insidious onset weakness of both lower limbs after she had undergone an obstetric procedure under spinal anesthesia. Patient was asymptomatic prior to this procedure. The patient presented to our tertiary care center, 2 months after the procedure, with progressive flaccid paraparesis (grade 2/5) and bowel and bladder involvement. MRI of the lumbo-sacral spine revealed a large septated cystic lesion, with the intensity of CSF on both T1WI and T2WI [Figure 5a], extending from the D10 vertebral level to the L5 vertebral level. The rim as well as the septations showed intense contrast enhancement [Figure 5b and c]. On the basis of a history of invasive procedure in the recent past, a working diagnosis of intradural spinal abscess was made. Unforunately, diffusion-weighted (DW) MRI was not performed at this time. The patient underwent L3-L4 hemi-laminectomy and drainage of the cystic lesion, but the cystic fluid revealed yellowish fluid. The cyst wall was sent for biopsy, which revealed schwannoma. The patient then underwent D11-L5 laminoplasty and total excision of the giant cystic schwannoma. She gradually recovered lower limb, bowel, and bladder function over a period of 3 months.

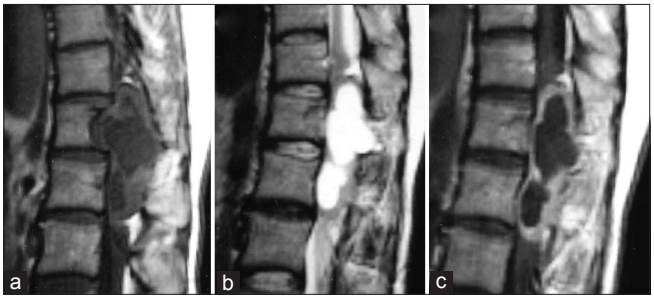

Figure 1: Sagittal MRI images of the dorso-lumbar junction spine (Case 1) showing multi-loculated cystic intradural extramedullary lesion at D11-L1 level. (a) Hypointense on T1WI; (b) Hyperintense on T2WI; (c) with intensely enhancing walls and septations on contrast injection

Table 1: Summary of clinical and radiological presentation and outcome of the patient group

Pt	Age/ sex	Clinical presentation	Type based on MRI findings	Surgery performed	Intraoperative findings	Post-operative recovery
1	35/M	Backache, progressive motor deficit below L1	Giant, cystic, multi-loculated [Figure 1]	D11-L1 laminectomy and total excision of lesion	Lobulated cystic lesion with thick walls; extradural extension+	Symptoms improved
2	45/F	Backache, focal neuro-deficit at L3-L4 level	Cystic, uniloculated [Figure 2]	L3-L4 laminectomy+total excision	Cystic lesion with translucent wall [Figure 2]	Power improved
3	45/M	Progressive motor deficit below L1 level	Cystic, multi-loculated [Figure 3]	D11-L1 hemilaminectomy and total excision	Multi-loculated thick walled cystic lesion with small solid component	Symptoms resolved
4	39/F	Spastic paraparesis	Cystic, uniloculated [Figure 4a]	D2-D3 Hemi-laminectomy and total excision	Cystic lesion with translucent walls	Spastic paraparesis improved
5	41/M	Spastic paraparesis	Cystic lesion loculations+at the lower end [Figure 4b]	D3-D5 Hemi-laminectomy+total excision	Cystic lesion with thick wall	Power improved and spasticity resolved
6	25/F	Flaccid paraparesis with bowel and bladder involvement	Giant, cystic multi-loculated [Figure 5]	D11-L5 Laminoplasty and total excision	Cystic lesion with tanslucent walls and septation	Partial improvement

MRI: Magnetic resonance imaging

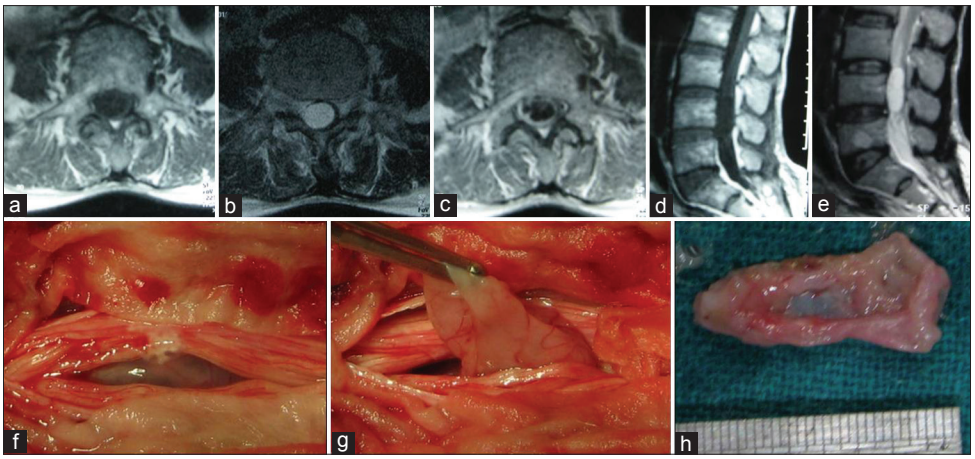

Figure 2: MRI imaging and intraoperaive findings in Case 2. (a,d) T1W axial and sagittal image showing hypointense lesion; (b,e) T2W axial and sagittal image showing hyperintense lesion; (c) Contrast-enhanced T1W axial showing thin rim enhancement; characteristic finding of uniloculated cystic spinal schwannoma. (f,g) Intraoperative findings showing cystic lesion with translucent, thin wall. (h) Specimen *ex vivo*

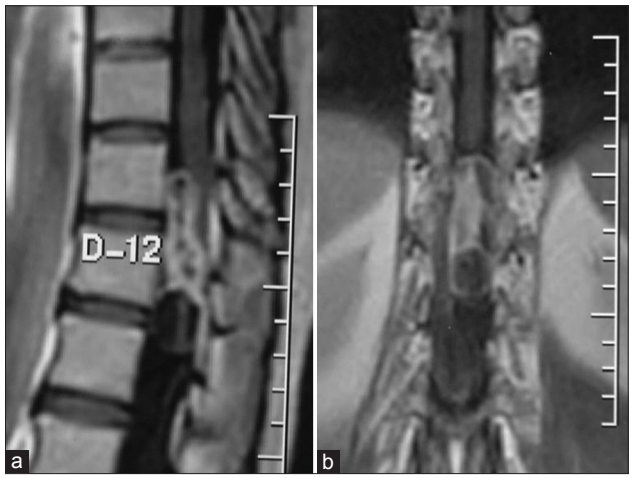

Figure 3: Contrast-enhanced Sagittal (a) and Coronal (b) MRI imaging findings in Case 3, showing a thick, irregular-walled, multiloculated cystic schwannoma

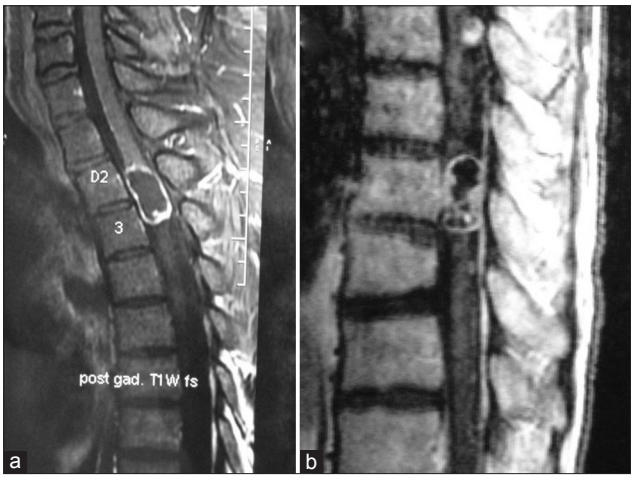

Figure 4: (a) Contrast-enhanced Sagittal MRI imaging in Case 4, showing uniloculated cystic spinal schwannoma at D2-D3 level with thin rim enhancement. (b) Contrast-enhanced Sagittal MRI imaging in Case 5, showing cystic schwannoma with loculations at the lower end

DISCUSSION

Schwannomas are the most common primary intraspinal tumors, accounting for approximately one-third of cases, usually occurring as solitary, well-circumscribed, encapsulated, solid or heterogeneously solid, eccentrically located, intradural extramedullary lesions on spinal nerve roots lesions in the cervical and lumbar region.[2,7]

Schwannomas are usually entirely solid or heterogenously solid tumors.[2] Predominantly cystic spinal schwannomas are uncommon lesions and may pose a preoperative diagnostic dilemma.[5] Various theories have been proposed to explain the cystic changes occurring in schwannomas. Degeneration of the Antoni B portion of a schwannoma can result in cyst formation, which may then progress to form a larger cyst.[5,6] Central ischemic necrosis/hemorrhage can be caused by tumor growth resulting in cyst formation within the tumor.[5,6] Another theory attributes cystic change in schwannomas to mucinous degeneration.[4,6] The first hypothesis likely explains the formation of totally cystic (uniloculated) schwannomas as seen in cases 2, 4, and 5 in our short series; while the other theories may hold good for the multiloculated cystic schwannomas, as seen in cases 1, 3, and 6 in our series.

The peak incidence of spinal schwannomas is in the fourth and fifth decades of life and they do not exhibit any predilection to a particular sex. There were six patients in our series of spinal cystic schwannomas; three males and three females. The age of the patients ranged from 25 to 45 years. The location of the cystic schwannomas was thoracic in two patients, thoraco-lumbar junction in two patients and lumbar in two patients. Patients harboring thoracic lesions presented with myelopathy in the form of spastic paraparesis, while patients with thoraco-lumbar junction and lumbar lesions presented with backache associated with symptoms of radiculopathy. Cystic tumors

have a high risk of causing progressive symptomatic worsening as a result of cyst expansion.[4] As seen in case 6 of our series, intrathecal spinal injection for anesthesia resulted in manifestation of the latent lesion.

MRI is the preferred imaging modality for establishing diagnosis. Schwannomas generally have low-to-intermediate signal intensity on T1WI. On T2WI, they may be heterogenous with focal areas of hyper- or hypointensity. Focal areas of intense hyperintensity on T2WI often correspond to cystic portions, whereas hypointensity may represent hemorrhage, dense cellularity, or collagen deposition.[3]

The differential diagnoses for the intradural extramedullary spinal lesions in our short series were neurenteric cyst, arachnoid cyst, hydatid cyst, epidermoid cyst, and dermoid cyst. The characteristic imaging findings for these differential diagnoses are detailed in Table 2.[1,3,5] In our series, a preoperative diagnosis of spinal cystic schwannoma was accurately made in only four out of the six patients (cases 1, 3, 4, and 5); in view of irregular character of the rim-enhancement in the lesions and multi-loculations within them. The walls of the lesions in these cases were slightly thicker and more irregular as compared with what one would see in a classical neurenteric, arachnoid, epidermal, or hydatid cyst. These findings as seen in [Figures 1, 3, 4b, and 5] should be definitely considered while evaluating such cystic lesions in this location. Imaging in case 2 suggested a preoperative diagnosis of arachnoid or hydatid cyst; however,

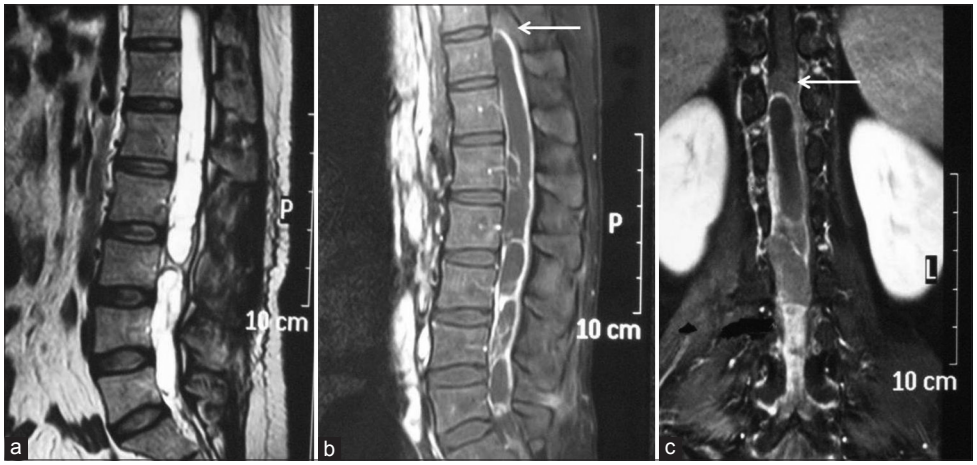

Figure 5: MRI findings in case 6. T2W Sagittal (a); contrast-enhanced T1W Sagittal (b); and contrast-enhanced T1W Coronal (c) MR images showing giant multi-loculated cystic schwannoma with a thin solid component along its cranial aspect (arrows). Preoperative differentials were intradural spinal abscess and cystic schwannoma. Intraoperative findings and histopathology confirmed cystic schwannoma

Table 2: Differential diagnosis of cystic spinal schwannomas: Analysis on MRI findings[1,3,5]

Lesion	Salient features	T1WI	T2WI	Contrast enhanced T1WI
Cystic schwannoma	Dorsal or lateral to spinal cord. Extradural extension with dumb-bell shape	Iso- to hypointense	Hyperintense	Intense rim enhancement+. Irregular walls+Septations+
Arachnoid cyst	Mostly seen in the thoracic spine. Situated dorsal to cord No restriction on DWI	Hypointense Smooth wall	Hyperintense Smooth wall	No contrast enhancement
Epidermoid cyst	Mostly seen in the lumbar spine. Commonly associated with spinal dysraphism. Restricted diffusion on DWI	Iso- to Hypointense on T1WI. (Slightly hyperintense to CSF)	Hyperintense	Nil to very mild enhancement. Spontaneous rupture with surrounding inflammation, may result in enhancing pattern
Dermoid cyst	Contents resemble that of fat. Scalloping of vertebral bodies+associated with dermal sinus	Hyperintense Fat suppression +	Hypointense	No enhancement
Neurenteric cyst	Ventral to cord. CT may show remnant through vertebral body. Anterior spina bifida is hallmark	Hypointense	Hyperintense	No contrast enhancement; Unless infected (rare)
Hydatid cyst	May be anywhere. Mostly multiple. Internal echoes+Daughter cyst+	Hypointense	Hyperintense	No contrast enhancement; unless complicated in the form of rupture or infection
Intradural spinal abscess	Lumbar spine is common location. Central Diffusion Restriction.	Hypointense	Hyperintense	Smooth, peripheral ring enhancing lesion

T1WI: T1-weighted MR image, T2WI: T2-weighted MR image, DWI: Diffusion-weighted imaging, CSF: Cerebrospinal fluid, MRI: Magnetic resonance imaging

retrospective analysis of the imaging revealed that the thin rim enhancement exhibited by the cyst wall should have been considered and a cystic schwannoma should have considered above the diagnosis of an arachnoid or hydatid cyst, which will exhibit rim enhancement only if complicated with rupture or infection. Clinical and imaging findings in case 6 suggested an intradural spinal abscess; however, intraoperative findings and histopathology confirmed the diagnosis of cystic spinal schwannoma.

The treatment of cystic schwannomas involves total excision of the lesion. This excision is recommended because inadequate removal has a risk of recurrence. In many large series, it has been confirmed that recurrence occurred in all cases in which the excision was subtotal.[2] Hemi-laminectomy (in the hands of experienced surgeons) or laminectomy at the level of lesion, followed by total excision was followed in treating these lesions at our institute, and good postoperative outcome was achieved in all patients.

CONCLUSION

Cystic spinal schwannoma are uncommon lesions, presenting as intradural extramedullary lesions, mainly in the dorso-lumbar spine. Contrast MRI is the investigation of choice and plays a major role in predicting these lesions preoperatively. Thick, irregular walls with septations, which intensely enhance on contrast injection, can accurately predict cystic multiloculated schwannomas. Uniloculated cystic schwannomas can be considered in cases of purely cystic lesions with rim enhancement of the thin wall. However, it is unlikely that all such cases can be predicted preoperatively on radiology. Frozen section histopathology should be used to identify them at surgery. It is important to detect these lesions at surgery, as total excision is possible and almost always results in good long-term neurological outcome.

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