



BMH Med. J. 2020;7(2):27-32. **Case Reports**

## **Cystic Schwannoma of the Spine – A Review of Literature and a Report of a Series of 5 Cases**

Suresh S Pillai, M Harisankar

Division Of Spine Surgery - Dept. Of Orthopedics, Baby Memorial Hospital Calicut, Kerala, India

**Address for Correspondence:** Dr. Suresh S Pillai, MBBS, D. Ortho, MS (Ortho), DNB (Ortho), MNAMS, Spine Fellow (Chang-Gung University, Taiwan), AO Spine Fellow (Nottingham, UK), MCh (Ortho), Consultant Spine Surgeon, Baby Memorial Hospital, Kozhikode, Kerala, India. PIN: 673004. E- mail: sureshorth@googlemail.com

### **Abstract**

Nerve sheath tumors are among the most common intradural spinal tumors, but intraspinal cystic nerve sheath tumors are a rare entity. MR imaging forms the diagnostic modality of choice. We discuss here 5 cases of cystic schwannoma of the spine, of which 4 were at the thoracolumbar junction (D12-L1), and one at D11-D12 level. The patients presented with varying degrees of cord compression symptoms including paresthesia, weakness in the lower limbs and in one patient with paraplegia. The treatment and prognosis of the cystic schwannoma lesion is basically similar to that of the solid schwannomas and includes complete excision of the tumor to prevent recurrence. Ki 67 index provides a reliable prognostic indicator to predict recurrence of a cystic schwannoma after surgical resection. All the patients were treated with en masse excision of tumor. The tumor tissue was assessed for Ki67 index, an index of tumor doubling time, and a reliable indicator of tumor recurrence in subtotal or incomplete removal. All the patients showed improvement in neurological status in the postoperative period. All the patients are on regular follow up and there has been no signs of recurrence yet.

**Keywords:** Nerve sheath tumors, cystic schwannoma, diagnosis, treatment, prognosis, Ki 67 index

### **Introduction**

Nerve sheath tumors are among the most common intradural spinal tumors [1], but intraspinal cystic nerve sheath tumors are a rare entity with only a handful of cases reported in the literature (eight cases according to Parmar et al) [1]. Even when a cystic lesion is detected in MR imaging, the possibility of a cystic nerve sheath tumors as a diagnosis is often not considered as schwannomas and neurofibromas are usually solid tumors [2]. Here we describe our experience with five intradural cystic schwannoma cases.

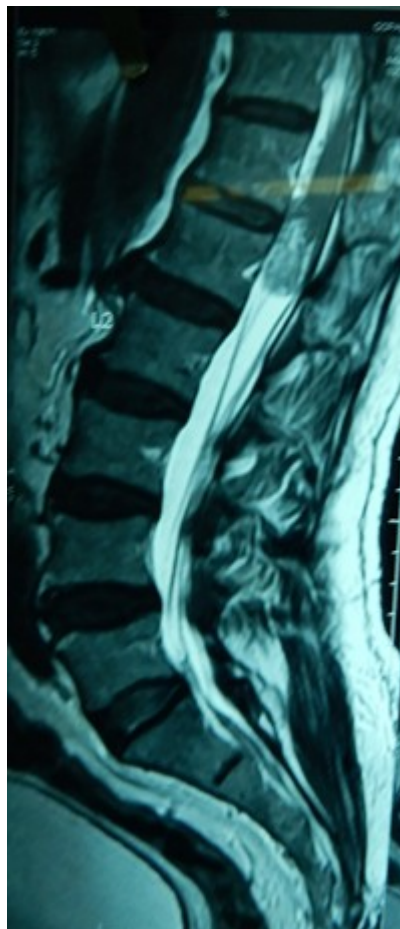
### Case reports

We discuss here 5 cases of cystic schwannoma of the spine, of which 4 were at the thoracolumbar junction (D12-L1), and one at D11-D12 level. The patients presented with varying degrees of cord compression symptoms including paresthesia, weakness in the lower limbs and in one patient with paraplegia.

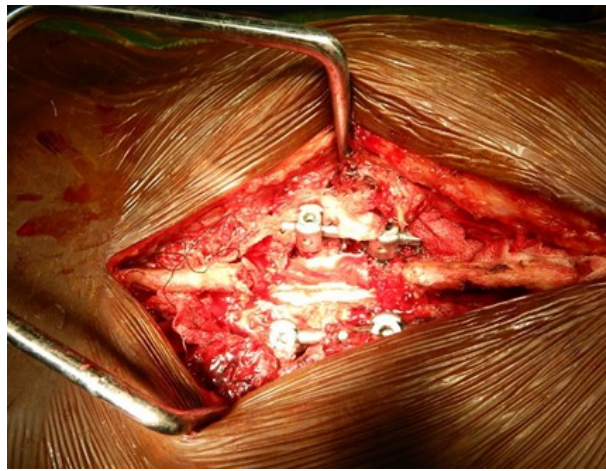
All the patients were treated with posterior instrumentation followed by laminectomy. The dura was opened, and the tumor identified. The tumor was isolated under microscopic examination and was resected en masse. The tissue in all the cases contained cystic lesion filled with clear fluid. The tissue was sent for biopsy and immunochemistry assay. The tumor tissue was assessed for Ki67 index, an index of tumor doubling time, and a reliable indicator of tumor recurrence in subtotal or incomplete removal [3,4]. All the cases showed Ki 67 index below 2 %, which is associated with very low chance of tumor recurrence [3]. The dural leak was closed, with 6-0 prolene. The dural closure was ensured to be watertight by doing intraoperative Valsalva maneuver.

All the patients showed improvement in neurological status in the postoperative period. All the patients are on regular follow up and there has been no signs of recurrence yet.

### Case 1



**Figure 1:** D12-L1 cystic schwannoma



**Figure 2:** Intraoperative picture showing instrumented vertebra and the intradural tumor.



**Figure 3:** Cystic schwannoma resected specimen

**Case 2**



**Figure 4:** Cystic schwannoma at D12- L1 level

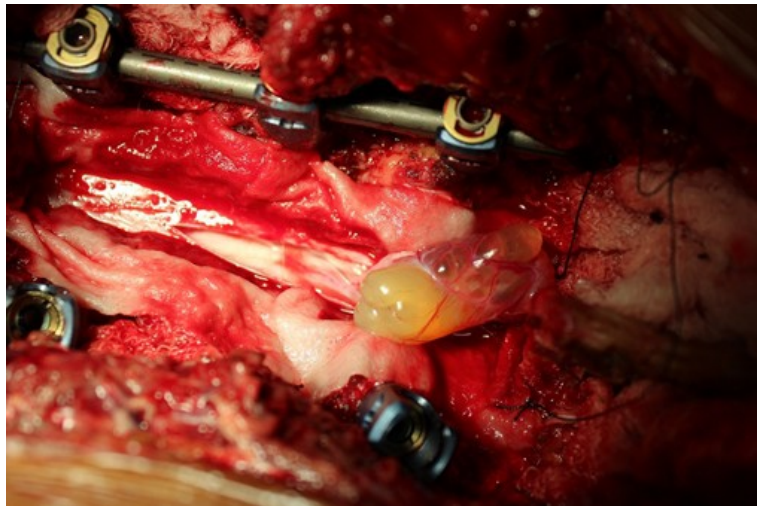


Figure 5: Intraoperative picture showing cystic schwannoma



Figure 6: Resected specimen of cystic schwannoma

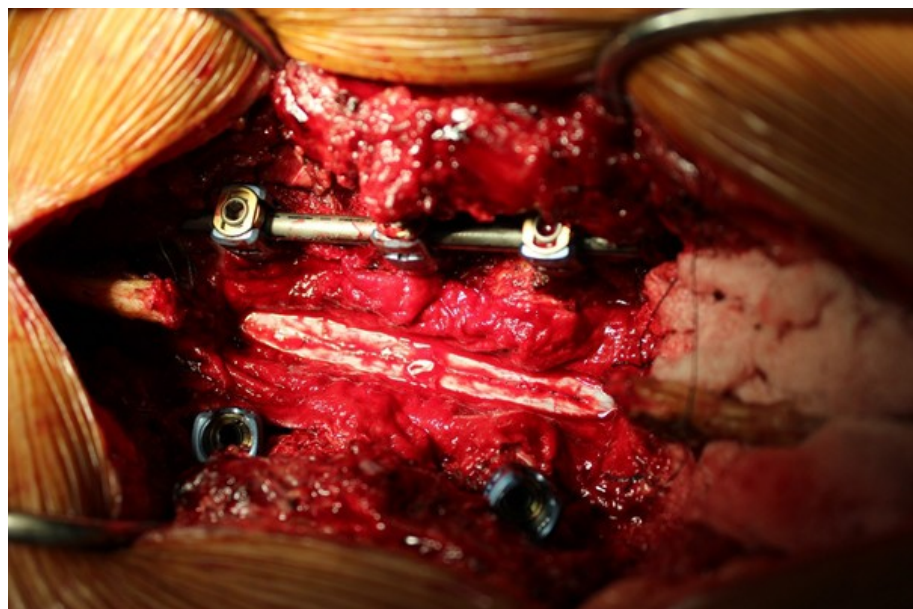


Figure 7: Dura after dural repair

## Discussion

Most common nerve sheath tumors in the spine are neurofibromas and cystic schwannomas. They account for 30 % of all spinal tumors [5]. Neurofibromas are usually found as a fusiform enlargement of the spinal nerve or found enveloping the dorsal sensory nerve roots. The histology reveals a peripheral myxomatous tissue and a

central fibrocollagenous tissue interspersed with schwann cells [2]. Meanwhile schwannomas arise from the embryonic neural crest cells and display a solitary nature, and are well circumscribed and encapsulated tumors. These are usually found eccentrically located on peripheral nerves or spinal nerve roots [1]. Histologically, they show areas of high cellularity (Antoni A) and areas of low cellularity (Antoni B) [2]. Schwannomas are the most common benign nerve sheath tumors. The lumbar region is the most common site for occurrence of spinal schwannomas [6]. Apart from lumbar region, these tumors have a predilection for lower cervical and thoracolumbar junction, while upper thoracic region is relatively spared [7].

Schwannomas present usually in the fourth and fifth decade of life with symptoms of root compression or spinal cord compression. Based on the location of tumor, this may even cause cauda equina syndrome [6].

Cystic degeneration in the substance of a solid schwannoma leads to the formation of a cystic schwannoma. Various theories have been proposed to explain this cystic changes. Degeneration of the Antoni B portion of the schwannoma is the most widely accepted theory [2,8,9]. The cyst most often contains serous or colourless fluid. When it has blood, and if it fails to clot, this gives rise to fluid – fluid levels in MR imaging [2].

MR imaging forms the main stay of diagnosis. Focal areas of hyperintensity within the normal low to intermediate signal intensity lesion (on T1 weighted imaging) indicate cystic changes. The other tumors that give a similar presentation are dermoid cysts, Tarlov cysts, arachnoid cyst, neurenteric cyst, epidermoid cyst and cystic teratoma [1,6].

The treatment and prognosis of cystic schwannoma is similar to that of solid schwannoma and includes complete excision to avoid recurrence of the tumor [2,6]. There is a 5 % chance for recurrence of a surgically treated schwannoma of the spine [10]. The chance of recurrence is assessed by assessing the Ki 67 index, which is a measure of tumor doubling time. Based on the level of Ki 67 index, patients are divided into two groups, Ki 67 index > 2% and Ki 67 Index < 2 %. Patients with index < 2% are associated with very low recurrence rates [3,4].

## Conclusion

When encountering a case of intradural cystic mass, a differential diagnosis of cystic schwannoma should be entertained. MR imaging forms the diagnostic modality of choice. The treatment and prognosis of the cystic schwannoma lesion is basically similar to that of the solid schwannomas and includes complete excision of the tumor to prevent recurrence. Ki 67 index provides a reliable prognostic indicator to predict recurrence of a cystic schwannoma after surgical resection. In the thoracic region, the rootlets associated with the lesion can be sacrificed without the risk of any sensory deficits as there is significant overlap between the sensory dermatomes in the thoracic region.

## References

1. Parmar H, Patkar D, Gadani S, Shah J. Cystic lumbar nerve sheath tumours: MRI features in five patients. *Australasian radiology*. 2001;45(2):123–127.
2. Coleman JF. *Robbins and Cotran's Pathologic Basis of Disease*. LWW; 2010.
3. Niemczyk K, Vaneecloo FM, Lecomte MH, Lejeune JP, Lemaitre L, Skarzynski H,

Vincent C, Dubrulle F. Correlation between Ki-67 index and some clinical aspects of acoustic neuromas (vestibular schwannomas). *Otolaryngology–Head and Neck Surgery*. 2000 Dec;123(6):779-83.

4. C Yokoyama M, Matsuda M, Nakasu S, Nakajima M, Handa J. Clinical significance of Ki-67 staining index in acoustic neurinoma. *Neurologia medico-chirurgica*. 1996;36(10):698-703.

5. Conti P, Pansini G, Mouchaty H, Capuano C, Conti R. Spinal neurinomas: retrospective analysis and long-term outcome of 179 consecutively operated cases and review of the literature. *Surgical neurology*. 2004;61(1):34-43.

6. Kumar S, Gupta R, Handa A, Sinha R. Totally cystic intradural schwannoma in thoracic region. *Asian journal of neurosurgery*. 2017;12(1):131.

7. Seppälä MT, Haltia MJ, Sankila RJ, Jääskeläinen JE, Heiskanen O. Long-term outcome after removal of spinal schwannoma: a clinicopathological study of 187 cases. *Journal of neurosurgery*. 1995;83(4):621–626.

8. Shiono T, Yoshikawa K, Iwasaki N. Huge lumbar spinal cystic neurinomas with unusual MR findings. *American journal of neuroradiology*. 1995 Apr 1;16(4):881-2.

9. Enzinger FM, Weiss SW. *Soft tissue tumors*. LWW; 1989.

10. Fehlings MG, Nater A, Zamorano JJ, Tetreault LA, Varga PP, Gokaslan ZL, et al. Risk Factors for Recurrence of Surgically Treated Conventional Spinal Schwannomas: Analysis of 169 Patients From a Multicenter International Database. *Spine*. 2016;41(5):390-398.