Spinal Intramedullary Schwannoma of the Conus

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Abstract

Schwannomas are one of the most common primary spinal tumors representing 30% of all intraspinal lesions. Intramedullary schwannomas constitutes 0.3% of all intraspinal tumors and approximately 1% of spinal cord schwannomas. Majority of the tumors are seen in the cervical (58%), followed by thoracic (32%) and lumbar (10%) regions. Very few are reported at the level of conus medullaris. The important radiologic features of intramedullary schwannoma are predominant extramedullary component, intramedullary spinal tumor with a thickened and enhancing spinal nerve root, absent syrinx, enhancing well with contrast, and sharp margins. The aim of surgery in intramedullary spinal schwannomas is total removal whenever possible. Diagnosing intramedullary schwannoma preoperatively needs high index of suspicion. We are reporting the 10th case of intramedullary schwannoma in the conus region.

Keywords

- Schwannoma
- Intramedullary
- Conus Medullaris

Introduction

Schwannomas are the tumors developing from Schwann cells of the nerve sheath.[1] Schwannomas are among the most frequent primary spinal tumors representing 30% of all intraspinal lesions. Most of the tumors are intradural extramedullary in location, but they also occur as purely extradural (25%) and dumbbell type (15%) in a proportion of cases. Intramedullary location of these tumors seems to be rare.[2] Intramedullary schwannomas constitute 0.3% of intraspinal tumors and approximately 1% of spinal cord schwannomas.[3] They have a slight preponderance for males (1.4:1) with a mean age at demonstration of approximately 49 years and it is reported to occur in as early as 9 years of age. The average duration of symptoms is 28 months, owing to the slow-growing nature of the tumor.[3] The majority of the tumors are seen in the cervical (58%), followed by lumbar (10%) and thoracic (32%) regions.[4] Very few are seen to be occurring at the conus medullaris. Only nine cases are reported in the conus region so far in the literature.[5] We are reporting an interesting case of intramedullary schwannoma in a middle-aged female in the conus region, the 10th case in the literature (►**Table 1**).

Case Report

A 40 years old female patient without any comorbidities was admitted with complaint of low back pain radiating to the left lower limb. On examination, she had normal power in all four limbs with intact sensation and normal bladder and bowel function. There were no neurocutaneous markers. She was evaluated with magnetic resonance imaging (MRI) lumbosacral spine which revealed a well-defined T1 and T2 hyperintense lesion with peripheral rim type of contrast enhancement at the level of L1 body extending to the upper part of L2 body with the expansion of the cord (►**Fig. 1**). There was no extramedullary component or thickening of the nerve root. With the preoperative diagnosis of ependymoma (the most common lesion in that region), the patient had a "D12–L2" laminectomy. Dura was opened in midline. There was slight discoloration noted on the left side of the cord. Midline myelotomy was done and an attempt was made to create the

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Table 1 Reported case in literature[5]

Serial No.	Year of reporting	Author	Age/sex
1	1967	Guidetti	M
2	1972	Bharathi	M
3	1982	Cantore	57/M
4	1983	Young	33/F
5	1983	Lesoin	28/M
6	1992	Jacquet	44/M
7	1995	Duong	53/F
8	1998	Hejazi	65/M
9	2009	Hayashi	78/F
10	2022	Present case	40/F

Abbreviations: F, female; M, male.

plane between the tumor and the cord on all sides, which was unsuccessful on the right and inferior portions. A tumour capsule was incised and internal debulking was done. The lesion was grayish in color, moderately vascular, non-suckable, and appears to be infiltrating the substance of the cord. Subtotal excision of the lesion was done leaving behind the portion infiltrating the cord. Postoperatively, patient was extubated on the table, with normal ankle and toe movements. The perianal sensation was intact with normal anal sphincter tone and bladder function. Histopathological examination revealed a moderately cellular tumor comprising spindle-shaped cells organized in interlacing fascicles (Antoni A) admixed with small foci of hypocellular areas (Antoni B). Occasional verocay bodies were seen showing nuclear palisading with intervening eosinophilic cytoplasm (►**Fig. 2**). There were thickened and hyalinized blood vessels with focal areas of perivascular lymphocytic infiltration. Mitosis and necrosis were absent, suggestive of intramedullary schwannoma. Immunohistochemistry showed strong positivity for S-100 (►**Fig. 2**). Postop MRI showed a residual capsule with a significant reduction in tumor volume (►**Fig. 2**). Patient was advised to be in regular follow-up.

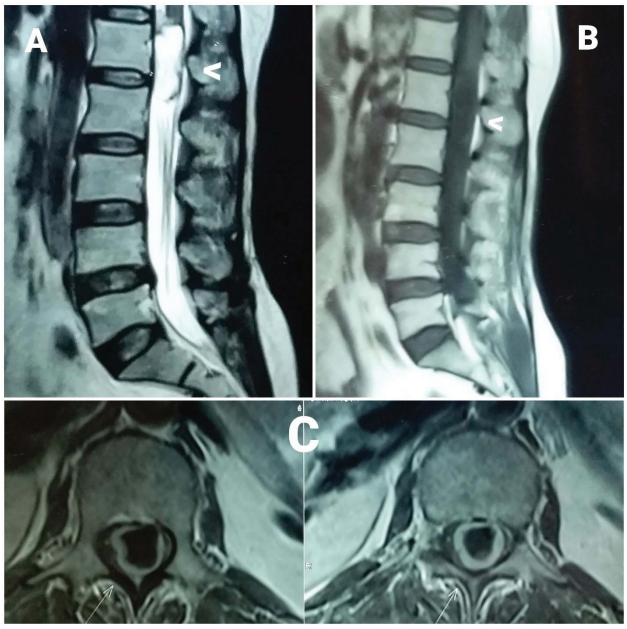

Fig. 1 (**A**) Well-encapsulated T2 hyperintense lesion at L1-L2 level. (**B**) Arrow head showing iso- and hypointense lesion noted in T1 image at L1-L2 level. (**C**) Axial postcontrast images showing intramedullary nature of the lesion

Discussion

Pathogenesis

Intramedullary schwannomas are very rare tumors, as the brain and spine lack Schwann cells and they are myelinated by oligodendrocytes. Though not clear, there are various hypotheses that describe the pathogenesis of intramedullary schwannomas.[6] They are:

1. Ectopic Schwann Cells Originating From Embryonic Neural Ridge.
2. The abnormal intramedullary nerve fibers ensheathed by Schwann cells.
3. Extension of Schwann cells along the anterior spinal artery's branches.
4. Schwann cell's neoplastically grow into the cord occurs at the sites in which the posterior nerve roots join the pia mater.
5. Pial cells of neuroectodermal origin undergo transformation into Schwann cells.

Diagnosis

The gold standard investigation to evaluate spinal intramedullary tumors is the MRI. If the intramedullary tumor is not associated with syringomyelia, then the diagnosis is more in favor of intramedullary schwannomas.[3] The important radiologic features[7] which probably suggest schwannoma are:

- ➢ Predominant extramedullary component.
- ➢ Intramedullary spinal tumor with a thickened and improving spinal nerve root.
- ➢ Absent syrinx.
- ➢ Intense contrast enhancement.
- ➢ Sharp margins.

Except for sharp margins and absent syrinx, other findings were lacking in our case.

Treatment

The surgical goal for intramedullary spinal schwannomas is total removal, whenever possible. Surgical excision of intramedullary schwannomas differs widely from extramedullary schwannomas. However, total excision may not be possible in all the cases, due to the adherence of the tumor to the surrounding neural tissue,[8] as in our case. Reoperation should be considered in case of recurence.[9] Adjuvant therapy like radiotherapy is not contemplated considering the benign nature of the lesion.

Conclusion

To conclude, diagnosing intramedullary schwannoma preoperatively needs a high index of suspicion with some

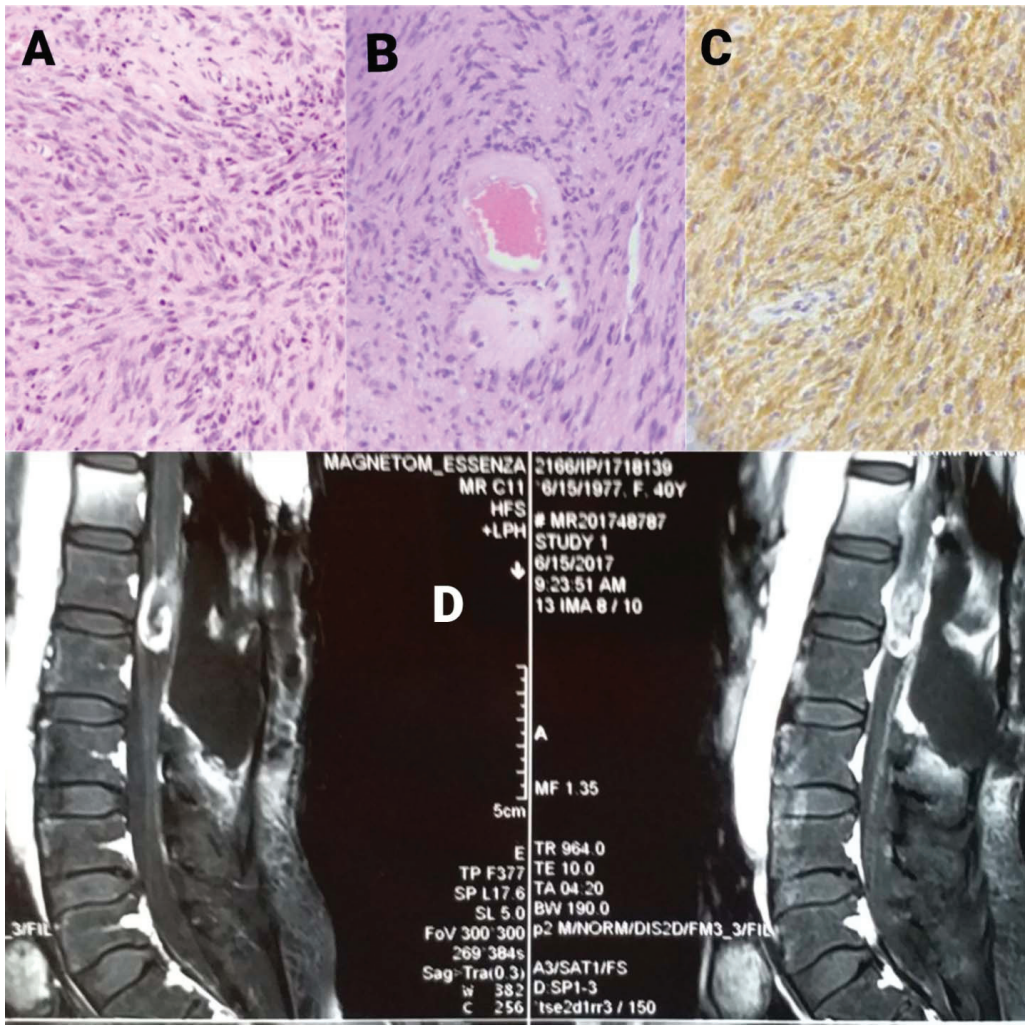

Fig. 2 (**A**) Microscopic picture showing hypercellular tumor (Antoni A) consisting of spindle-shaped cells. (**B**) Microscopic picture showing verocay body and hyalinized blood vessel. (**C**) Immunohistochemistry showing strongly and diffuse positivity for S-100. (**D**) Postoperative scan showing enhancing residual capsule.

features suggesting it. The extent of resection needs to be decided intraoperatively, depending on the adherence/infiltration of the tumor to the substance of the spinal cord. Patients who underwent partial excision of the tumor have to be followed up for recurrence. Reoperation should be considered for patients developing recurrence.

Conflict of Interest
None declared.

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