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Case Report

Intramedullary schwannoma – A case report

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ABSTRACT

Background: Schwannomas are benign but clinically progressive tumors. Mostly, they present as intradural extramedullary lesions. They are quite rare in the intramedullary (IM) region. We report a case of IM schwannoma.

Case Description: A 52-year-old gentleman presented with a history of gait instability and numbness in bilateral lower limbs. He had clinical signs of myelopathy. His magnetic resonance imaging (MRI) dorsal spine was done that showed an intradural IM lesion at the level of D11, with one differential of ependymoma. Near total resection of lesion was done and histopathology reported it schwannoma.

Conclusion: Preoperative radiologic assessment for IM spinal lesions is difficult and high degree of suspicion should be present when approaching a patient with somatic pain and IM lesion on MRI, keeping in mind one differential of IM schwannoma.

Keywords: Gross total resection, Histopathology, Intramedullary schwannoma

INTRODUCTION

Schwannomas account for 30% of primary intraspinal tumors. They are mostly intradural extramedullary, with only 1% of these reported in the literature as intramedullary (IM).[5]

Schwannomas usually arises from the dorsal sensory roots. Patients with extramedullary schwannoma commonly present with pain. IM Schwannomas, on the other hand, can present with wide range of symptoms. Few of these include pain, dysesthesias, and pyramidal syndrome followed by sensitivity complaints and sphincter dysfunction.[5,11]

Majority of cases of IM schwannoma have been reported in the cervical spine (63%), followed by thoracic (26%) and lumbar segments (11%) of spinal cord.[5]

Schwannomas originate mainly from the Schwann cells of peripheral nerve sheaths in the spinal canal and hence are rare within the spinal cord which lacks Schwann cells. The pathogenesis of IM schwannoma is still unclear and various hypothesis (listed below) has been devised that can lead to development of IM schwannoma.

- Schwann cells along the IM perivascular nervous plexus
- Ectopic Schwann cells that originate from migrating neural crest cells
- Schwann cells related to aberrant IM myelin fibers
- Focal IM proliferation of Schwann cells in reaction to chronic diseases or trauma.[10,12]

IM schwannomas are picked up on magnetic resonance imaging (MRI), and their definitive diagnosis is confirmed on biopsy following surgical excision. Since they are quite rare in the literature, we report a case of IM schwannoma in a 52-year-old gentleman that was diagnosed based on MRI, followed by surgical excision and histopathology. With the advancement in MRI and preference for operative management of all IM lesions, IM schwannomas are likely to be described more often in coming years.[7]

CASE REPORT

We report a case study of 52-year-old gentleman with no known prior comorbids, who presented to neurosurgery clinic with complaints of frequent instability in gait and numbness in bilateral lower limbs, for around 3–4 weeks. Recently, his symptoms have worsened to an extent that he is only able to walk with support, or else he falls within 5–6 steps.

At presentation, his neurological examination revealed powers of 5/5 in both upper limbs, 5/5 in the lower limbs proximally and 4 + distally. Signs of myelopathy including brisk lower limb reflexes and bilateral upgoing plantars were noted. Sphincters were continent and no sensory impairment was appreciated.

His MRI dorsolumbar spine with contrast was performed which reported an IM abnormal signal intensity area with homogenous enhancement, measuring 1.4*1.5*2.1 cm, corresponding to the level of T11, causing compression of spinal cord anteriorly and to the left side. This is associated with high signal cord edema in the lower thoracic spinal cord. No evidence of dural tail and extension in exiting foramina was noted. The lesion appears isointense on T1 and iso to hypointense on T2, with post contrast-enhancement; findings representing IM neoplasm with one differential of ependymoma [Figure 1].

With the impression of an IM cord tumor, patient was planned for surgical excision. After informed consent and preoperative counseling, D10 partial, D11 complete, and D12 partial laminectomy were performed.

Intraoperatively, an IM lesion was noted with no definite arachnoid planes, myelotomy was performed by the tumor itself. Hence, near total resection of tumor was done. Neuromonitoring could not be used because of unavailability.

Postoperatively, the patient had no new sensory or motor deficits in the lower limb. His active physiotherapy and limb strengthening exercises were done.

Histopathology of biopsied lesion reported areas of compact, elongated cells, with occasional nuclear palisading (Antoni A pattern) and less cellular, loosely textured cells with indistinct processes and variable lipidization (Antoni B pattern). Nuclear palisading and fibrillary processes were seen in cellular areas. Immunohistochemical stain S100 was positive in neoplastic lesion and GFAP negative in tumor cells. These findings were consistent with the diagnosis of schwannoma [Figures 2 and 3].

DISCUSSION

This report presents a case of 52-year-old gentleman with IM schwannoma, which is quite rare in the IM region, due to absence of Schwann cells in central nervous system. The last possible literature review was done in 2018 that quoted 70 cases of IM schwannoma reported by then.[5]

IM schwannomas present commonly with somatic pain and other neurologic deficits are mostly manifested later.[1] MRI is the preferred imaging modality for diagnosis. IM schwannoma most commonly appears as iso to hypointense on T1W MRI and hyerintense on T2W images.[3,11] However, variations in radiological signal intensity of tumor do occur.[2] These differences are due to the macroscopic variants of tumor, that is, cystic versus solid, and also type of cells seen on biopsy, that is, Antoni A versus B.[9]

Preoperative radiologic diagnosis of IM schwannoma, based on MRI alone, is challenging.[5,12] IM tumor with extramedullary component can be confidently diagnosed as schwannoma. However, differentiation of purely IM lesion from other types of tumors based on radiology alone is often practically impossible.[6] Misdiagnosis of IM schwannoma as glioma may lead to unsuitable treatment. Therefore, thorough radiological and clinical assessment, along with a high index of suspicion, should always raise one differential of IM schwannoma in mind, despite their low incidence.[5] Occasionally, intraoperative differentiation of IM tumors is also difficult and requires the use of frozen section analysis to clear the diagnostic dilemma.[7]

Unlike the more malignant lesions of cord such as glioma and schwannomas are usually benign and have well defined cleavage plane.[8] This makes total resection achievable in most cases, offers the best clinical outcome, and avoids subsequent recurrence.[1,8] Hence, the preferred mode of treatment for IM schwannoma is gross total surgical resection to an extent as much as possible, without causing any neurologic deficit.[5] However, in certain infiltrative variants of IM schwannoma, this may not be possible.[11] When the tumor is adherent to neural tissue, sub-total resection is likely to improve neurological function followed by a second surgery later or adjuvant radiotherapy. The role of adjuvant radiotherapy is not well established.[5]

The accurate diagnosis of tumor is based on histopathology. To have good functional status, surgery should be performed timely before any permanent neurologic deficit develops.[13]

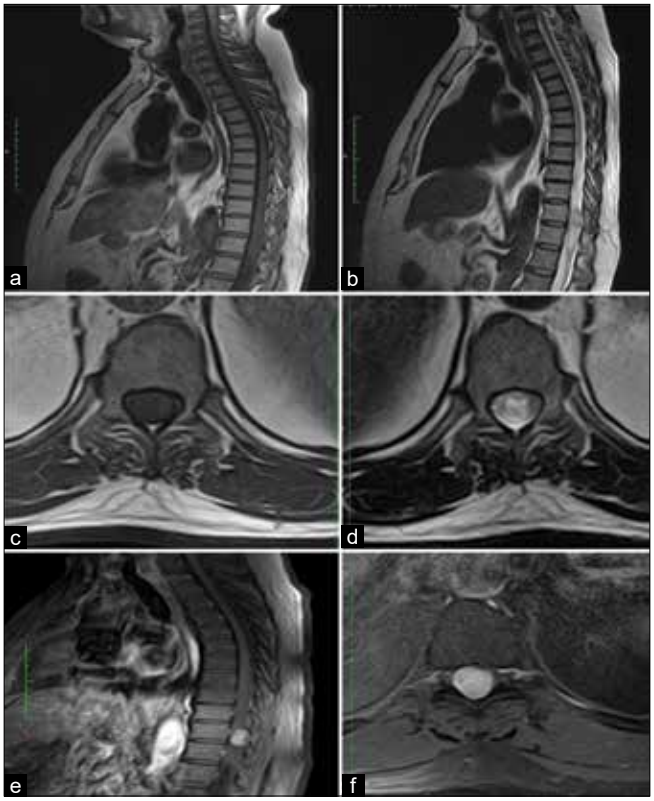

Figure 1: (a and b) (Sagittal T1- and T2-weighted images), (c and d) (Axial T1- and T2-weighted images), (e and f) (Contrast-enhanced sagittal and axial T1-weighted images) showing IM lesion which is isointense on T1, slightly hyperintense on T2 and shows post contrast-enhancement.

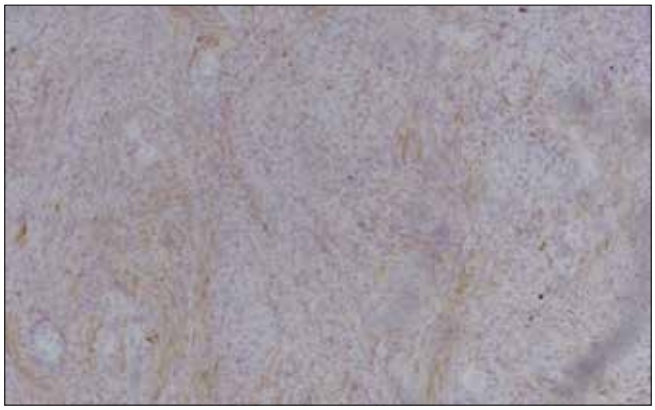

Figure 2: IHC stain GFAP is negative in tumor cells.

Conventional schwannomas mostly have a good prognosis, with <5% recurrence reported in the literature. However, data on long-term follow-up of IM schwannoma are lacking. Dhake and Chatterjee reported recurrence in two cases of thoracic IM schwannoma, highlighting the need for long-term follow-up in this group of patients.[4]

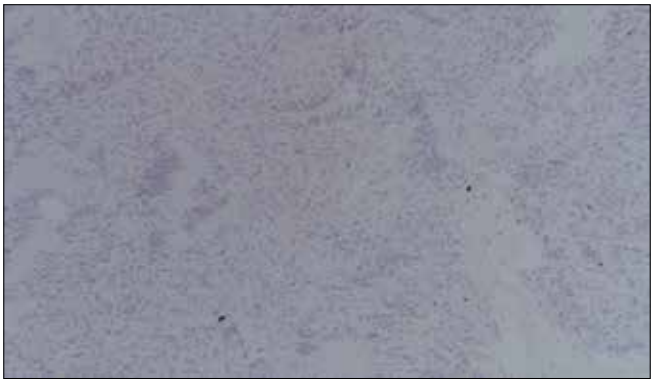

Figure 3: IHC stain S-100 is positive in tumor cells.

CONCLUSION

IM schwannomas are difficult to be picked up on radiologic and clinical assessment preoperatively. Since misdiagnosis may lead to inappropriate treatment[5] and the definitive diagnosis of these lesions can only be made on histopathology, a high degree of suspicion for such lesions should always be kept in patients presenting with chronic somatic pains, dysesthesias without any signs of severe neural damage; and IM lesions on MRI. These lesions can be treated with gross total resection and good prognosis can be guaranteed in majority of such patients.[5,8]

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest

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