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Case Report

Intra Medullary Schwannoma - Case Report

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Abstract

Background: Schwannoma's are benign but clinically progressive tumors. Mostly, they present as intradural extramedullary lesions. They are quite rare in the intramedullary region. We report a case of Intramedullary Schwannoma.

Case presentation: Fifty two year-old gentleman presented with history of gait instability and numbness in bilateral lower limbs. He had clinical signs of myelopathy. His MRI dorsal spine was done, that showed an intradural intramedullary 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 intramedullary spinal lesions is difficult and high degree of suspicion should be present when approaching a patient with somatic pain and intramedullary lesion on MRI, keeping in mind one differential of intramedullary schwannoma.

Keywords: Intramedullary (IM); Schwannoma; Gross total resection; Histopathology

Introduction

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

Schwannoma's usually arise from the dorsal sensory roots. Patients with extramedullary schwannoma commonly present with pain. Intramedullary Schwannomas on the other hand, can present with wide range of symptoms. Few of these include pain, dysesthesias, pyramidal syndrome followed by sensitivity complaints, and sphincter dysfunction [1,2].

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 [1].

Schwannoma's 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 intramedullary schwannoma is still unclear and various hypothesis (listed below) have been devised that can lead to development of intramedullary schwannoma.

- Schwann cells along the intramedullary perivascular nervous plexus.
- Ectopic Schwann cells that originate from migrating neural crest cells.
- Schwann cells related to aberrant intramedullary myelin fibers.
- Focal intramedullary proliferation of Schwann cells in reaction to chronic diseases or trauma [2-4].

Intramedullary schwannomas are picked up on MRI, and their definitive diagnosis is confirmed on biopsy following surgical excision. Since they are quite rare in literature, we report a case of intramedullary 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 intramedullary lesions, IM schwannoma's are likely to be described more often in coming years [5].

Case Presentation

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 to 6 steps.

At presentation, his neurological examination revealed powers of 5/5 in Both Upper limbs, 5/5 in 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 intramedullary abnormal signal intensity area with homogenous enhancement, measuring 1.4 cm × 1.5 cm × 2.1 cm, corresponding to the level of T11, causing compression of spinal cord anteriorly and to 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 intramedullary neoplasm with one differential of ependymoma (Figure 1).

With the impression of an intramedullary cord tumor, patient was planned for surgical excision. After informed consent and pre-operative counselling, D10 Partial, D11 complete and D12 Partial Laminectomy was performed.

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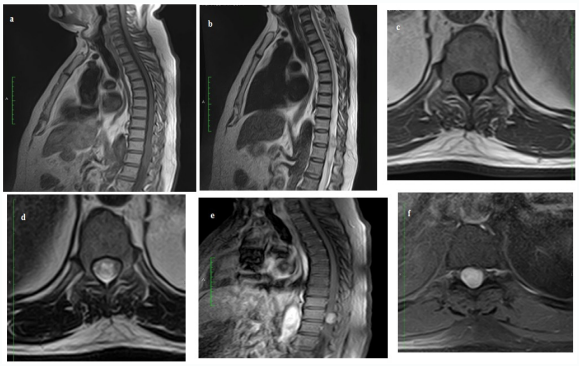

Figure 1: (a, b): Sagittal T1 and T2 weighted images, (c, d): Axial T1 and T2 weighted images, (e, f): Contrast enhanced sagittal and axial T1 weighted images. Showing intramedullary lesion which is isointense on T1, slightly hyperintense on T2 and shows post contrast enhancement.

Intraoperatively, an intramedullary 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.

Post operatively, 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 intramedullary Schwannoma, which is quite rare in the intramedullary region, owing to absence of Schwann cells in CNS. The last possible literature review was done in 2018 that quoted 70 cases of IM schwannoma reported by then [1].

Intramedullary schwannomas present commonly with somatic pain and other neurologic deficits are mostly manifested later [6]. MRI is the preferred imaging modality for diagnosis. Intra medullary schwannoma most commonly appears as iso to hypo-intense on T1W MRI and hyperintense on T2W Images [2,7]. However, variations in

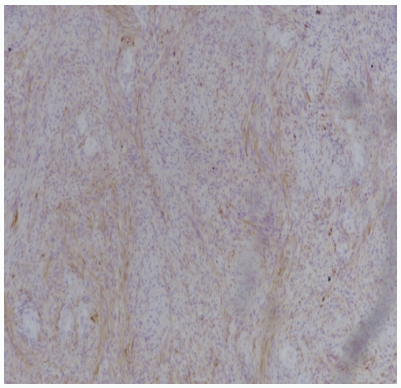

Figure 2: IHC stain GFAP is negative in tumor cells.

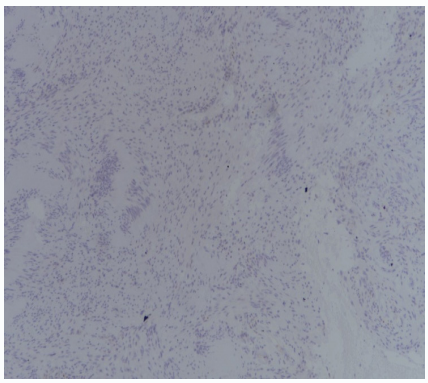

Figure 3: IHC stain S-100 is positive in tumor cells.

radiological signal intensity of tumor do occur [8]. These differences are due to the macroscopic variants of tumor i.e. cystic *vs.* solid, and also type of cells seen on biopsy i.e. Antoni A *vs.* B [9].

Pre-operative radiologic diagnosis of IM schwannoma, based on MRI alone, is challenging [1,4]. Intramedullary tumor with extramedullary component can be confidently diagnosed as schwannoma. But differentitation of purely intramedullary lesion from other types of tumors based on radiology alone is often practically impossible [10]. Misdiagnosis of intramedullary 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 intramedullary schwannoma in mind, despite their low incidence [1]. Occasionally, intraoperative differentiation of IM tumors is also difficult and requires the use of frozen section analysis to clear the diagnostic dilemma [5].

Unlike the more malignant lesions of cord like glioma, schwannomas are usually benign and have well defined cleavage plane [11]. This makes total resection achievable in most cases, offers the best clinical outcome and avoids subsequent recurrence [6,11]. 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 [1]. However, in certain infiltrative variants of intramedullary schwannoma, this may not be possible [2]. 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 radio therapy is not well established [1].

The accurate diagnosis of tumor is based on histopathology. In order to have good functional status, surgery should be performed timely before any permanent neurologic deficit develops [12].

Conventional Schwannomas mostly have a good prognosis, with less than 5% recurrence reported in literature. However, data on long term follow up of intramedullary schwannoma is lacking. Dhake et al. [13] reported recurrence in 2 cases of thoracic intramedullary schwannoma, highlighting the need for long term follow up in this group of patients.

Conclusion

Intra medullary schwannomas are difficult to be picked up on radiologic and clinical assessment preoperatively. Since misdiagnosis may lead to inappropriate treatment, and the definitive diagnosis of these lesions can only be made on histopathology, a high degree of

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suspicion for such lesions should always be kept in patients presenting with chronic somatic pains, dysesthesia without any signs of severe neural damage; and intramedullary lesions on MRI. These lesions can be treated with gross total resection and good prognosis can be guaranteed in majority of such patients.

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