Intramedullary meningioma of spinal cord: Case report of a rare tumor highlighting the differential diagnosis of spinal intramedullary neoplasms

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Abstract

A 15-year-old male presented with progressive weakness of both lower limbs with urinary incontinence. Magnetic resonance imaging revealed a spinal intramedullary mass at D7-D8 level. The child was operated with a preliminary diagnosis of an intramedullary tumor. Atypical ependymoma and astrocytoma were considered in the differential diagnosis. Per- and post-operative histopathological examination reported the case as transitional meningioma (WHO Grade I). Spinal intramedullary meningiomas being a rare entity may be confused with other common intramedullary tumors. Though, rare still the possibility of an intramedullary spinal mass of being a meningioma does exist and therefore should be considered in the differential diagnosis of intramedullary tumors.

How to cite this article:

How to cite this URL:

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INTRODUCTION

Meningiomas are among the most common spinal tumors, constituting about 25% of all adult intraspinal tumors. Spinal meningiomas are commonly encountered in the thoracic segments of the spine, usually in the subdural and extradural regions. Intramedullary location is extremely rare for these tumors, and so far to the best of our knowledge only seven case reports have appeared in the literature. [1] Modern neuroradiological tools have made the diagnosis of intramedullary masses easier and allow careful surgical planning. However, the presentation of meningioma as an intramedullary mass poses a preoperative diagnostic challenge. This case is discussed here for its exceptional location and to highlight the fact that spinal meningiomas may occur in locations other than extra/intradural and therefore should be considered in the differential diagnosis of spinal intramedullary masses.

CASE REPORT

A 15-year-old male presented with complaints of progressive weakness of both lower limbs for 4 months. Weakness was insidious in onset and associated with stiffness of lower limbs. He also complained of moderate intensity low backache with a 1-month history of bladder incontinence.

On neurological examination, the patient showed decreased strength in both lower limbs with hyper-reflexia and spasticity. The patient showed sensory loss of 60% below the umbilicus for touch, temperature, and pain.

Magnetic resonance imaging (MRI) of the dorsal spine was performed. Multiplanar T1, T2-weighted (T1-W and T2-W) and post contrast fat suppressed T1-W sequences were obtained. The study showed focal expansion of the mid dorsal cord effacing subarachnoid space with altered intramedullary signal intensity and craniocaudal cord edema [Figure 1a]. Post contrast T1-W images showed an oval well-defined intensely enhancing ×12 mm at the D7-D8 level [Figure 1b and c]. A provisional diagnosis of intramedullary spinal cord tumor was made with atypical ependymoma and astrocytoma considered as the differentials. [Figure 2]

The patient subsequently underwent a surgery. A laminectomy was done at D6-D8 level revealing dural bulge. A midline durotomy was followed by a midline myelotomy between the dorsal columns. The tumor was encountered after the columns were split. Intraoperative findings revealed a soft to firm, greyish white, moderately vascular, and nonsuckable mass. A clear plane was identifiable between cord and tumor. Subtotal resection was done with a peroperative diagnosis of ependymoma. The per-operative specimen sent to the histopathology laboratory was subjected to squash smear preparation and frozen sections. It was difficult to crush and spread. The squash smear stained by hematoxylin and eosin showed occasional whorls and numerous single uniform spindled meningothelial cells [Figure 2a]. Frozen section showed whorl formation by the meningothelial cells [Figure 2b]. Based on the above findings, a per-operative histopathological diagnosis of meningioma was made. [Figure 2]

The latter specimen received in formalin for paraffin sections showed whorls and fascicules of meningothelial cells having ovoid nuclei with stippled chromatin and eosinophilic cytoplasm [Figure 2c]. Tumor cells showed immunopositivity for vimentin [Figure 3a] and immunonegativity for glial fibrillary acidic protein [Figure 3b]. Histopathological diagnosis of transitional meningioma (WHO Grade I) was made. The postoperative period was uneventful.

The histopathological diagnosis of intramedullary transitional meningioma (WHO Grade I) led to a reconsideration of the original radiological diagnoses, the MRI scans were studied retrospectively, and a diagnosis of intramedullary meningioma was established. The 2-year follow-up has showed considerable improvement in the patient status. [Figure 3]
Most spinal meningiomas are confined within the intradural space and only a few of them penetrate the dura or exit through a root sleeve to reach the epidural compartment. [2]

Infrequently, meningiomas appear to arise in the epidural compartment or the skin [3],[4] and very rarely are spinal meningiomas intramedullary. Pathogenesis of intramedullary meningioma is not very clear, but as per the hypothesis the origin of these tumors is in the mesenchymal cells lining the perivascular spaces of the neuraxis. [5],[6]

This case report highlights the differential diagnosis of common spinal intramedullary neoplasms so as to render a definitive diagnosis.

Spinal intramedullary meningiomas must be distinguished from a group of neoplasms, more frequent at this location. This group includes astrocytic neoplasms, ependymomas, oligodendrogliomas, ganglion cell tumors, haemangioblastomas, paragangliomas, schwannomas, and metastatic neoplasms.

Astrocytomas of the spinal cord are broadly divided into two clinicopathologically distinct groups: (i) Diffuse or diffusely infiltrative astrocytomas and (ii) Pilocytic astrocytoma. Combined approach of neurosurgery, radiology, and pathology modalities ensures an accurate assessment of the lesion. Spinal astrocytomas range from Grade II to IV. These occur in both children and adults presenting with pain, motor deficits, sensory disturbances, and abnormalities of sphincter function. MRI revealing an ill-defined nonenhancing expansile mass extending over several segments characterizes Grade II or III astrocytoma while an ill-defined contrast enhancing mass is characteristic of Grade IV astrocytoma. The radiologic features of pilocytic astrocytomas are yet to be described completely; however, in a study of Rossitch et al., this tumor is defined as discrete, cyst associated, and contrast enhancing mass. [7]

Spinal ependymomas are the commonest gliomas of the spinal cord and are divided into (i) cellular ependymoma and (ii) Myxopapillary ependymoma. Cellular ependymomas are common during the 4th and 5th decade. These may be multifocal in the setting of NF2. [8] MRI features of ependymomas show discrete and contrast enhancing masses. T2-W images may show dark hemosiderin rich regions. [8] Associated peritumoral cysts are also a common feature.

Per-operatively the discrete character and central location of the tumor is usually apparent. The tumors are usually solid but can be cystic also. Despite of these features the diagnosis is sometimes difficult in frozen sections since the cytologic features of ependymomas may simulate astrocytomas. In such situations, smears complement the frozen sections highlighting the small dark oval ependymal nuclei.

Myxopapillary ependymoma is a distinctive neoplasm seen in adults. Clinical features include sphincter disturbances or deficits related to compression/involvement of conus medullaris or sacral nerve roots. MRI shows discrete and contrast enhancing masses. The characteristic abundant mucin shows hyperintensity or whiteness on nonenhanced T1-W images. Macroscopically these are well-defined, soft, saccular masses that may secondarily adhere to the nerve roots of cauda equina or extend to involve the conus medullaris. The crush smears show the typical mucoid matrix with cells showing nuclear uniformity and process formation.

Spinal haemangioblastomas are seen in adults during 3rd -5th decades, predominantly in men. This tumor shows an occasional association with von Hippel Lindau disease. MRI shows a discrete, contrast enhancing mass with associated cysts, syringes above or below the mass. Macroscopically the high vascularity of the lesion results in compensatory enlargement of leptomeningeal vessels simulating a vascular malformation at times. The cut surface of the tumor has a distinctive variegated appearance due to the lipidization. In frozen sections, haemangioblastomas often resemble astrocytomas. The cellular crowding and the occasional nuclear pleomorphism accentuated by the freezing process can create the impression of even anaplastic astrocytoma (WHO Grade III). However, review of radiologic studies and the vacuolated appearance of lipid containing stromal cells on frozen sections help in clinching the diagnosis. In addition, a positive lipid stain on the frozen tissue confirms the diagnosis.

Paragangliomas are uncommon lesions, commonly seen in adults. MRI shows discrete and contrast enhancing mass. The characteristic Zellballen architecture and distinctive nuclei of chief cells with a "salt and pepper" chromatin pattern are well seen in cytologic preparations.

Oligodendrogliomas and Ganglion cell tumors both are rare but recognized entities in the spinal cord showing features similar to their cranial counterparts. Similarly, intramedullary schwannomas and metastatic neoplasms are uncommon lesions which need a reminder that they may appear at this site.

The other intramedullary neoplasms those are too rare to affect the spinal cord includes: Germinoma, a typical teratoid or rhabdoid tumor, hamartoma, lipoma, lymphomatoid granulomatosis, neurocytoma, primary CNS lymphoma, primitive neuroectodermal tumor and teratoma.

**CONCLUSION**

Intramedullary meningioma is a very rare clinical entity. Owing to their rarity, the imaging characteristics of intramedullary meningiomas have not been well characterized. However, when confronting a homogenously enhanced intramedullary tumor of the cord on postcontrast MRI, meningioma should be considered in the differential diagnoses.

**References**