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Case Report

Long segment cervicothoracic intramedullary dermoid with concomitant conal lesion – A case report

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ABSTRACT

Background: Spinal dermoid cysts are benign tumors that result from congenital or acquired ectodermal inclusions. Long segment intramedullary involvement of the spinal cord is exceedingly rare, and there are only a handful of case reports found in the literature.

Case Description: A 30-year-old female presented with a 3-month history of myelopathy characterized by progressive quadriparesis and urinary incontinence. Magnetic resonance imaging revealed multifocal heterogeneous intramedullary masses extending from C2 to T4 and at T12–L1 with similar intensity lesions seen within the central cord from T5 to T11 level. Following tumor decompression, she showed significant improvement in neurological function 1 month later. The histopathological examination confirmed the diagnosis of a multifocal intramedullary dermoid cyst.

Conclusion: Partial surgical extirpation is a reasonable treatment for long segment intramedullary dermoid cysts, particularly when the tumor capsule is adherent to critical adjacent neural tissues.

Keywords: Dermoid cyst, Inclusion cyst, Intramedullary tumor, Spinal tumor

INTRODUCTION

Spinal dermoid cysts are benign tumors that result from congenital or acquired ectodermal inclusions.[5] Histologically, they are characterized by the presence of dermal structures. These tumors account for 0.8–1.1% of all primary spinal tumors with the majority occurring in the extramedullary or subdural juxtamedullary lumbosacral region (e.g., the conus or cauda equina).[4-6]

Here, we report a C2–L1 long segment cervical to conus medullaris intramedullary dermoid cyst along with satellite lesions.

CASE DESCRIPTION

History and examination

A 30-year-old female presented with a 3-month history of progressive quadriparesis and urinary incontinence. The neurological examination revealed atrophy of the intrinsic muscles of the

hands, mild bilateral triceps weakness, and bilateral lower extremity weakness (4/5). Deep tendon reflexes were absent bilaterally at the triceps levels but hyperactive in both lower extremities with bilateral Babinski responses.

Imaging

The MR revealed multifocal heterogeneous intramedullary masses extending from C2 to T4 and T12–L1; similar intensity lesions were seen within the central cord from T5 to T11 level [Figures 1-4]. Signal characteristics were inhomogeneous on both T1- and T2 weighted sequences. Fat-suppressed images were consistent with lipid content in all the lesions and there was no enhancement with contrast. Further, there was mild accompanying intramedullary edema surrounding the tumors.

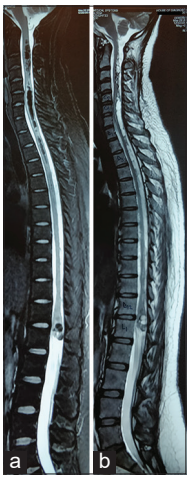

Figure 1: T2 weighted fat saturated (a) and T2 weighted (b) magnetic resonance imaging of the whole spine – sagittal view.

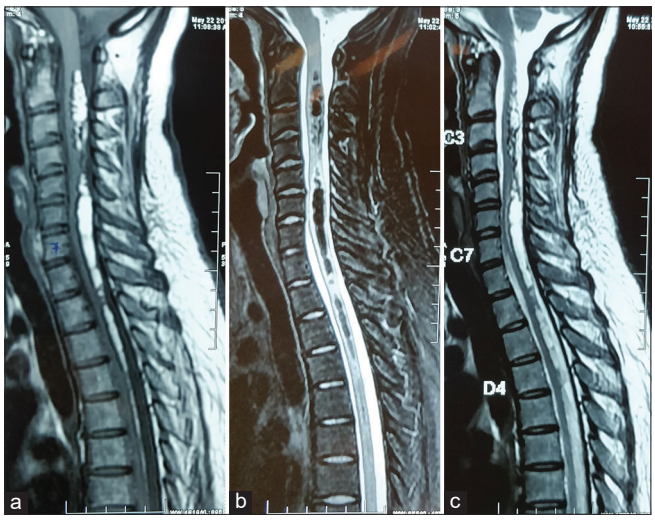

Figure 2: Sagittal views of the cervicodorsal spine in T1 (a), T2 fat saturated (b) and T2 weighted (c) magnetic resonance images.

Surgery

The patient underwent a C2–T4 and T12–L1 laminectomy utilizing intraoperative SSEP and MEP neuromonitoring. Once the dura mater was opened, an expanded spinal cord was immediately identified [Figure 5]. Following a midline myelotomy, partial resection of the intramedullary lesion was performed. The conus lesion was yellowish, waxy, friable, and contained with intermingled well-formed hair. Internal debulking was continued until we encountered an adherent capsule; at this point, a subtotal resection of the C2–T4 and T12–L1 lesions was performed (without resecting tumor at other levels) to avoid incurring increased neurological morbidity. Once the cord was decompressed, the dura was closed primarily. No additional laminoplasties were performed.

Postoperative course

The postoperative period was uneventful and the patient was discharged on postoperative day 6 requiring intermittent catheterization. Within 1 month, she reported improvement in all modalities. No further postoperative MR imaging was available.

Histopathology

Biopsies from multiple sites were suggestive of a dermoid cyst showing: stratified squamous epithelium along with fibro-collagenous tissue with blood vessels, glial tissue, hair shafts, adipocytes, and areas of calcification [Figure 6].

DISCUSSION

Frequency and epidemiology

Dermoid tumors are rare lesions which constitute around 0.8–1.1% of intraspinal tumors.[4-6] Although dermoid tumors develop during the embryonic period, they are slow-growing lesions and do not cause symptoms until the second or third decades of life. There is a slight male predominance.[6]

Imaging

Due to the varying amounts of soft tissue, fat, calcium, and hemorrhage, MRI typically demonstrates heterogeneous signal intensity lesions with the relatively high signal from fat (e.g., on MRI T1W images) making the identification of its lipid component more readily apparent.[5,6]

Rupture tendency for spinal dermoids

Spinal dermoid cysts are known to rupture, and their content can spread throughout the subarachnoid space and

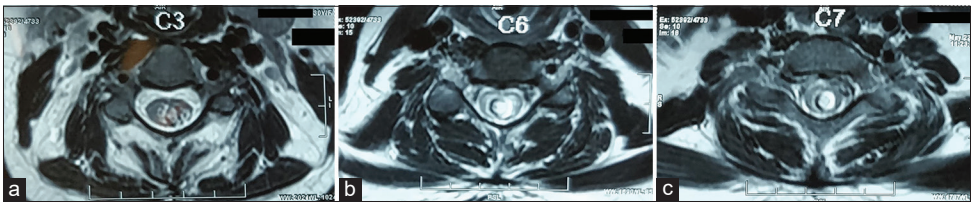

Figure 3: Axial cuts of T2 weighted magnetic resonance imaging showing heterogeneously intense intramedullary lesions within the cord at C3 (a), C6 (b) and C7 (c) vertebral levels.

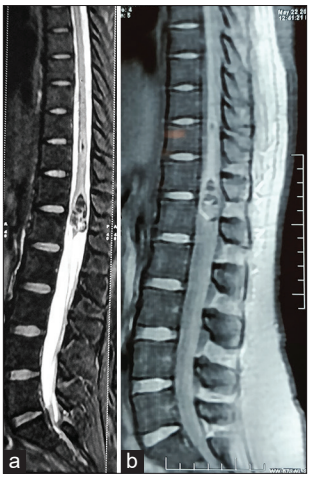

Figure 4: Sagittal views of the lumbosacral spine in T2 fat saturated magnetic resonance images without (a) and with (b) contrast.

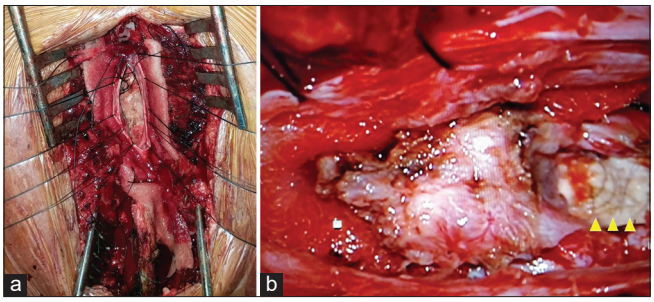

Figure 5: (a) Intraoperative photograph showing the intradural exposure of the conal lesion. (b) Visualization of the tumor under the operative microscope with well-formed hairs (yellow arrowheads) within the lesion.

ventricular system.[1,2] In our case, no rupture occurred. Although holocord abscesses secondary to infected dermoids have been reported, our case showed no evidence of meningismus and/or infection.[7]

Prior reports of extensive intramedullary dermoid cysts

A holocord intramedullary dermoid has only been reported once in the literature involving a patient who refused surgery.[5] Shukla *et al.* reported a long segment intramedullary dermoid from T4 to L1 vertebral level which was surgically debulked.[6]

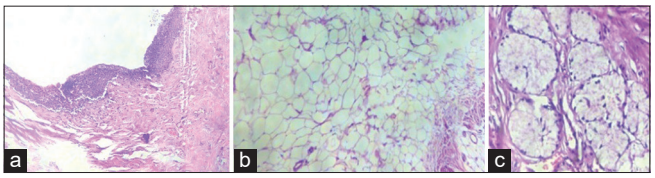

Figure 6: Histopathology microscopic images (H and E ×20) showing (a) cyst lined by stratified squamous epithelium, (b) mature adipose tissue, (c) mucinous glands.

Recommended treatment of intramedullary spinal dermoids

Given the indolent, benign nature of these tumors, asymptomatic lesions should ideally be managed conservatively.[4] Notably, if gross total radical excision is likely to incur a major neurological deficit (e.g., due to an adherent capsule to critical neural structures); then, biopsy, partial, or subtotal removal may be performed.[3,4] Further, for extensive intramedullary lesions, skip laminectomies may provide decompression at critical locals while providing representative histological confirmation of the tumor.

CONCLUSION

Intramedullary spinal dermoid cysts are rare tumors. Here, we report successful partial resection of intramedullary cord lesion extending from C2–L1 treated with skip laminectomies C2–T4 and T12–L1 laminectomies.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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