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DORSO-LUMBER INTRAMEDULLARY DERMOID CYST WITHOUT SPINAL DYSRAPHISM : A RARE ENTITY

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ABSTRACT Spinal dermoids are uncommon, especially in an intramedullary location. Absence of spinal dysraphism associated with spinal dermoid is extremely rare and with best of our knowledge only seven cases have been reported in the literature till date. This is a case of a 9-year-old boy, who presented with progressive weakness of both the lower limbs. Magnetic resonance imaging (MRI) showed an intramedullary lesion extending from D11 to L1 with peripheral enhancement on contrast. Decompression of the cystic contents with partial removal of cyst wall was done. Hair with sebum & fat was encountered. Histopathology confirmed the diagnosis of dermoid cyst. This case adds to the previous reported cases of the rare and uncommon intramedullary space occupying lesions of the spinal cord.

KEYWORDS : Spinal Dysraphism, Intramedullary Tumors, Intraspinal Dermoid Cysts

INTRODUCTION

Intramedullary dermoid cysts of the cord are rare, with very few cases been reported in the literature till date with absence of spinal dysraphism. [1] Intraspinal dermoid cysts are usually located in lumbar and thoracic regions and are usually associated with congenital spinal dysraphisms and or dermal sinus tracts.[2,3,4]

Absence of spinal dysraphism associated with spinal dermoid is extremely rare and with best of our knowledge only seven cases have been reported in the literature till date.[5] Our case is 9-year-old boy with intramedullary dermoid cyst in the dorso-lumbar cord without associated spinal dysraphism.

Case summary

A 9-year-old male, who presented with weakness of both the lower limbs (Right> Left), & gradual thinning of both the lower limbs (Right> Left), since six months with chief complaints of difficulty in walking, & unable to bear weight in lower limbs. There was also complaint of pain abdomen due to retention of urine & constipation since three months. On examination, there was wasting of both the lower limbs right side being more affected than the left. Power in right lower limb was Medical Research Council (MRC) grade 1 and left was MRC grade 2 with flaccidity, and decreased tone in both lower limbs. Patient was bed ridden since the last 2 months. Plantars were bilateral mute with absent ankle and knee jerks in both lower limbs. The skin over the neck and back was normal with no evidence of any sinus, hairy patch, or any cutaneous mark. MRI of the dorso-lumbar spine with contrast study showed widening of the spinal cord from D11 to L1 with peripheral ring enhancement **[Figure -1A].** There was hyper intensity in part of the lesion on T1W images, suggestive of fat component **[Figure -1B].** The lesion was hyper intense on T2W images **[Figure -1C] .**

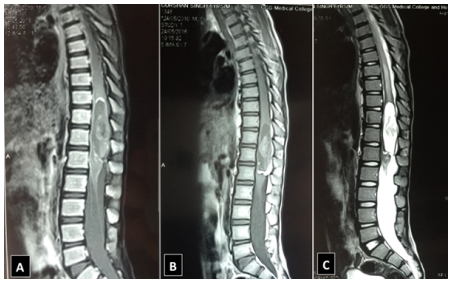

FIGURE 1 A:- T1W post-contrast image revealed peripheral ring enhancement and irregular enhancement within the lesion, **B-** T1W image revealed hyperintensity in posteroinferior part of intramedullary space occupying lesion suggestive of fat component, **C** T2W image showed a hyperintense lesion.

Patient was operated under general anesthesia in prone position. A midline incision extending from D10 to L2 was made. Para spinal muscles were retracted laterally by subperiostially dissection. D10 to L2 laminectomy was done. Intra operatively the dura was bulging and the cord was expanded. A midline myelotomy was made. Hair with grayish white shiny debris & fat was removed. The myelotomy was subsequently extended rostrally and caudally. Grayish white grumous contents mixed with hair & fat was removed and the cyst was decompressed **[Figure -2A].** Only some part of the cyst wall was removed. In major part of the cyst, the wall was densely adherent to the cord and attempt to remove that would have led to damage to the cord. Hence, it was left behind. The myelotomy was left open **[Figure -2B].**

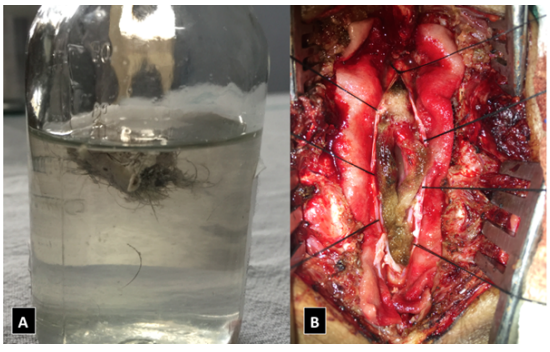

FIGURE 2 A:- Excised dermoid - grayish white contents mixed with hair & fat. **B** Operative cavity after excision of dermoid cyst

Immediate post op patient's power in the lower limbs was MRC grade 0. Over a period of three weeks, power improved to MRC grade 3 in the lower limbs. Catheter was kept for two months & then removed, and the patient was able to pass urine normally. Bowel symptoms also improved during that period. The patient was able to walk with support after three months of surgery.

Histopathological analysis confirmed the diagnosis of dermoid cyst

DISCUSSION

Spinal dermoid account for 0.8-1.1% of primary spinal tumors and the majority are in the extramedullary or subdural juxtamedullary in the lumbosacral region.[6] Many of the intraspinal dermoid cysts are seen

in association with other congenital anomalies including spina bifida occulta, dorsal dermal sinus, myelomeningocoele, tethered cord, hypertrichosis, and other dermal abnormalities. The association of intramedullary dermoid without spinal dysraphism is extremely uncommon. The dermoid cysts are developmental in origin. They arise from the embryonic ectoderm in the developing embryo.

The nervous system develops from the ectoderm. In the developing embryo, the cells on its dorsal aspect thicken to form neural plate. The neural tube bends and closes & from it the whole of the nervous system develops. The neural tube has the anterior neuropore & posterior neuropore which closes at 24 days and at 28 days respectively. The closure occurs in the dorsal midline first in the cervical region and then extending cranially and caudally. Therefore, in the lumbo-sacral i.e. the caudal part, the neural tube closes last & there is more chance that this process may be disturbed. Dermoid cyst arises from the nests of cutaneous tissue which gets trapped within the developing neural tube. Thus, in the spine, lumbo-sacral region is the most common region for dermoids.

Dermoids are very commonly associated with spinal dysraphisms as the process which gives rise dermoid cysts also causes spinal dysraphisms. The low incidence of dermoid in the cervical region is because the closure of neural tube begins in the area destined to become the lower cervical cord and proceeds rostrally and caudally. Spinal inclusion cysts are neuroenteric cysts, arachnoid cysts, epidermoid and dermoid cysts which are intradural, extramedullary in location. The most common time of presentation of dermoid cysts is childhood. The symptoms arise because of cord tethering and mass effects.

But, in the present case, the child had no associated developmental anomaly of the spine. The dermoid cyst was removed by micro neurosurgical technique as done for other intramedullary tumors. The capsule of the dermoid was very much adherent to the cord & it was not possible to remove it completely. Therefore, part of the capsule was left behind as an attempt to remove it totally would have caused unacceptable damage to the cord.

CONCLUSION

The intramedullary location of the dermoid cyst with the absence of any congenital spinal dysraphism makes this case a very unique and rare entity and adds to the reported cases of rare intramedullary space occupying lesions.

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