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CASE REPORT

Dermoid cyst: A rare intramedullary inclusion cyst

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ABSTRACT

Intramedullary dermoid cysts are rare tumors, especially those not associated with spinal dysraphism. Only six cases have been reported in the literature. Of these, only two cases have had magnetic resonance imaging studies. We report a case of an 18-year-old female patient, who presented with progressive weakness of both the lower limbs and wasting of both the upper limbs. Magnetic resonance imaging (MRI) showed an intramedullary lesion extending from C3 to D2 with peripheral enhancement on contrast. Decompression of the cystic contents with partial removal of cyst wall was done. Hair with oily cholesterol and keratin debris 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.

Key words: Intramedullary cysts, intramedullary tumors, intraspinal dermoid cysts

Introduction

Inclusion cysts of the cord are rarely intramedullary, with only few isolated cases been reported. Intraspinal dermoid cysts are usually located in lumbar and thoracic regions and are usually associated with congenital spinal dysraphisms and or dermal sinus tracts.[1-3]

Dermoid cysts within the cervical cord without associated spinal dysraphism are extremely rare with only six cases reported in the literature, till date,[4-9] and only two such cases with magnetic resonance imaging (MRI) studies. We report a case of intramedullary dermoid cyst in the cervical cord in an 18-year-old female patient.

Case Report

Presentation

Our patient was an 18-year-old female, who presented with gradual thinning of both the upper limbs (Left>Right) and weakness of both the lower limbs (Left>Right), since one and half years in the form of difficulty in walking and two episodes of fall.

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On examination, there was severe wasting of both the upper limbs left side being more affected than the right. Power in left lower limb was 4/5 and right was 5/5 with spasticity, and in the upper limb was 3/5 on left side and 4/5 on right side with weakness of hand grip. Patient was able to walk only with the help of a walker. Plantar were bilateral extensors with exaggerated ankle and knee jerks and ill sustained clonus of left patella and ankle. Upper limb reflexes were absent. There were no sensory changes in upper as well as lower limb, and in rest of the body, the skin over the neck and back was normal with no evidence of any sinus, hairy patch, or any cutaneous mark.

MRI of the cervical spine with contrast study showed widening of the cervical spinal cord from C3 to D2 [Figure 1]. An intramedullary space occupying lesion was present, which was hypo intense on T1W images, hyper intense on T2W images, and with peripheral ring enhancement on contrast study [Figures 2-4]. Also, "whorls" were seen within the lesion on T2 weighted images [Figure 4]. There was associated excavation of the bony spinal canal and the cervical vertebral bodies.

Operative technique

Patient was operated under general anesthesia in prone position. Injection of methyl-prednisolone was given intraoperatively and continued in the postoperative period for 23 hours at a dose of 30 mg/kg bolus over 1 hour followed by 5.4 mg/kg/hr for 23 hours. A midline incision extending from just below the external occipital protuberance to D3 was made. Para spinal muscles were retracted laterally by subperiostially dissection. Partial C2 and C3 to D3 laminectomy were done. After laminectomy, the cervical dura was found to be bulging and enlarged. Dura was opened in the midline and retracted by stay sutures. The cord was enlarged. A small midline

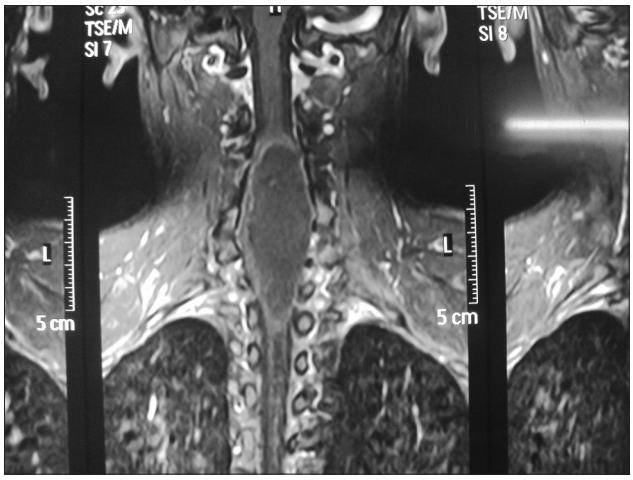

Figure 1: T1W coronal MRI showing widening of the cervical cord

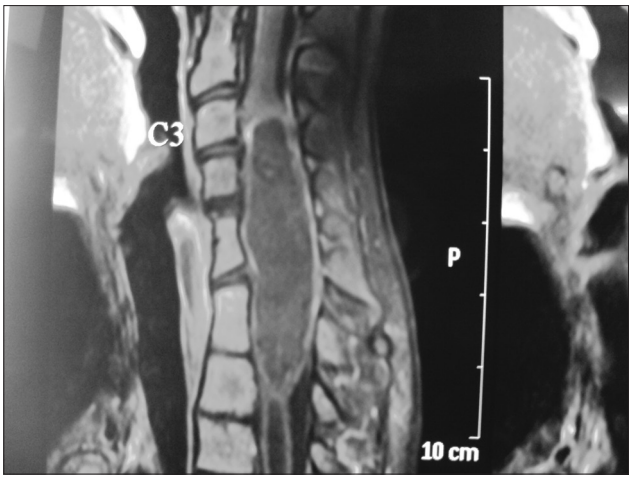

Figure 2: T1W image showing hypointense intramedullary space occupying lesion

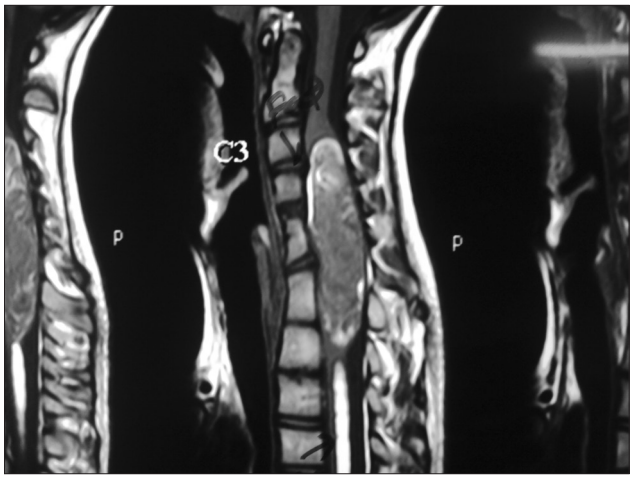

Figure 3: T1W post-contrast image showing peripheral ring enhancement and irregular enhancement within the lesion

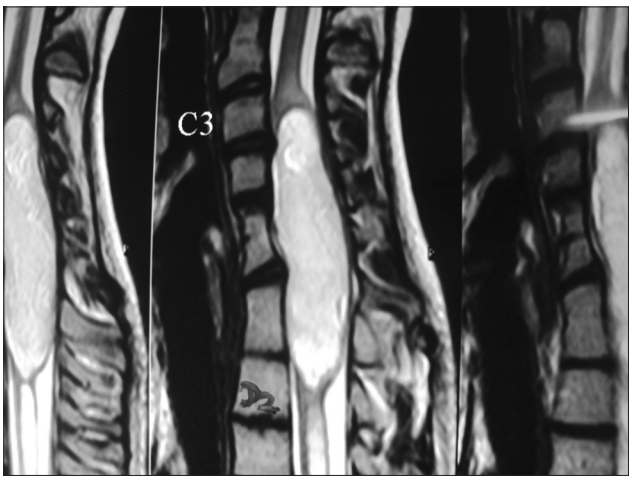

Figure 4: T2W image showing the hyperintense lesion with "whorls"

myelotomy was made initially. White shiny debris with hair was removed. The myelotomy was subsequently extended rostrally and caudally. Whitish grumous contents mixed with hair was removed and the cyst decompressed. Only some part of the cyst wall, which was not adhered densely to the cord, was removed. In major part of the cyst, the wall was densely adherent to the cord and attempt to remove them would have lead to damage to the cord. Hence, it was left behind.

The myelotomy was left open. Dura was closed with vicry 4-0. Muscles and skin were subsequently closed with absorbable stitches. Recovery from anesthesia was uneventful.

Results

Immediately after the surgery, patient's power in the lower limbs was 0/5 and 3/5 in the upper limbs. There was no respiratory distress. Over period of three days, power improved to 3/5 in the lower limbs and 4/5 in the upper limbs (Right>Left). Catheter was removed on 7th post operative day, and the patient was able to pass urine normally. Wound was healed and stitches were removed on 8th post-operative day. The patient was able to walk with support on the 10th post-operative day and was discharged.

Histopathological analysis confirmed the diagnosis of dermoid cyst.

Discussion

The common locations of dermoid cyst are

1. Scalp (angle of eye and retro mastoid region)
2. Skull bones (intradiploic)
3. Intracranial, in suprasellar region and posterior fossa
4. Intraspinal mainly intradural and associated with other spine defects.

The dermoid cysts are developmental in origin and arise from the nests of embryonic ectoderm which get buried or trapped under the lines of fusion of the ectodermal folds in the developing embryo.

The nervous system develops from the ectoderm. The cells on the dorsal aspect of the developing embryo thicken to form neural plate or placode along the axis of the embryo. The neural tube bends and closes to from a tube called the neural tube from which the whole of the nervous system develops. The neural tube closes in the dorsal midline first in the cervical region and the closure then extending cranially and caudally so that the anterior neuropore closes at 24 days and posterior neuropore at 28 days. Thus, as the neural tube closes last in the caudal part, that is the lumbo-sacral region, there is more chance that this process may be disturbed and nests of cutaneous tissue may get trapped within the developing tube, giving rise to dermoid cyst.

Hence, lumbo-sacral region is the most common site for the dermoids in the spine.

Also, dermoids are commonly associated with spinal dysraphisms. This is because the same process which gives rise to spinal dysraphisms is responsible for the development of dermoid cysts. The low incidence of dermoid in the cervical region is likely related to the embryological process of neural tube closure, which begins in the area of the neural tube destined to become the lower cervical cord and proceeds rostrally and caudally.

Spinal inclusion cysts are usually intradural, extramedullary in location, the common lesions being neuroenteric cysts, arachnoid cysts, epidermoid and dermoid cysts.

Dermoid cysts usually present themselves in childhood, as a consequence of associated anomalies or by symptoms of cord tethering and mass effects. However, in this case, the patient had no associated developmental anomaly of the spine. Because of the absence of any other congenital anomaly of the spine, the patient presented at a later age after she had developed significant symptoms, particularly in the left side of the body. The decompression of the dermoid cyst was carried out by standard micro neurosurgical technique employed for other intramedullary tumors.

However, it was not possible to remove the capsule of the dermoid completely as it was very much adherent to the cord. Any attempt to remove it totally would have lead to unacceptable damage to the cord; and hence, part of the capsule was left behind.

We could found two such cases[2,3] where a dermoid cyst was in the cervical cord and was not associated with any other congenital anomalies of the spine. In other reported cases,[3-7] the location of the dermoid cyst and the presence or absence of congenital anomaly of the spine is not clear as they have been published in language other than English and they are dated before the advent of MRI. Hence, it is difficult to ascertain whether these cases represent the "true" intramedullary dermoid cyst as in our case or are part of a developmental defect.

Conclusion

The intramedullary location of the dermoid cyst in the cervical cord and the absence of any congenital spinal dysraphism make this case a very unique and rare entity and add to the reported cases of rare intramedullary space occupying lesions.

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