Abstract: Intramedullary dermoid cysts without spinal dysraphism are very rare. Only seven cases have been reported in the literature till now. Of these, only three cases underwent Magnetic resonance imaging (MRI) 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 hand muscles. MRI showed an intramedullary lesion extending from C5 to C7 with peripheral contrast enhancement. Complete removal of lesion along with cyst wall was done. Hair with keratin debris, tooth and sebaceous glands were encountered. Histopathology confirmed the diagnosis of dermoid cyst.

Keyword: Intramedullary dermoid cyst, MRI, spinal dysraphism

Introduction

Inclusion cysts of the spinal cord are rarely intramedullary, with only few cases have been reported. Intraspinal dermoid cysts are commonly located in the lumbar and thoracic regions and are usually associated with congenital spinal dysraphism and dermal sinus. Intramedullary dermoid cyst in the cervical region without spinal dysraphism are extremely rare, only seven cases were reported in the literature till date. We report a case of intramedullary dermoid cyst in the cervical cord in an 18-year-old female patient.

Presentation

Our patient was an 18-year-old female, who presented with gradual thinning of the muscles of both hands, and weakness of both the lower limbs of three years duration in the form of difficulty in holding objects, mixing food and difficulty in walking. On examination, there was severe wasting of both thenar and hypothenar muscles, right hand being affected more than the left. Power in both lower limbs was 4/5 and with spasticity, and in the upper limb was 3/5 on right side and 4/5 on left side with weakness of hand grip (right-20%, left-30%). Patient was able to walk with difficulty. Plantars were extensor bilaterally with exaggerated ankle and knee jerks and sustained clonus of both ankle. In upper limb triceps, supinator reflexes were absent. There were no sensory changes in upper as well as lower limbs, and in rest of the body.

Cervical intramedullary dermoid cyst-a rare case presentation
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There was a hairy patch over the upper dorsal spines. MRI of the cervical spine with contrast study showed widening of the cervical spinal cord from C5 to C7 [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 Also, “whorls” were seen within the lesion on T2 weighted images [Figure-2]. There was associated excavation of the bony spinal canal and the cervical vertebral bodies.

Figure-1

Figure-2

Figure-3
Operative technique
Patient was operated under general anesthesia in prone position. Intraoperatively, injection of methyl-prednisolone was given 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 C2 to D1 was made. Para spinal muscles were retracted laterally by sub periosteal dissection. Partial laminectomy of C5, C6, C7 and D1 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 and a low grade torsion was noted. A small myelotomy was made in the root entry zone initially. About 1 to 1.5 ml of yellowish colored fluid drained. The myelotomy was subsequently extended rostrally and caudally. White shiny debris with hair [Figure-3] was removed. Whitish pultaceous contents mixed with hair and teeth [Figure-4] were removed. Complete excision of cyst with cyst wall done. The myelotomy was left open. Dura was closed with 6-0 prolene. Muscles and skin were subsequently closed with absorbable stitches. Recovery from anesthesia was uneventful.

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 abnormality 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 form 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 process which gives rise to spinal dysraphisms is also 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, extra medullary 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, small part of the capsule was left behind.

We could find three 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.

References