A Case of Thoracic Spine Intramedullary Dermoid Cyst
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Abstract: Intraspinal dermoid cysts are rare and benign congenital tumors that occur due to the defective closure of the neural tube. They can be intramedullary (rare) & extramedullary (common). They comprise 1% of intracranial and intraspinal tumors. Most common location is lumbosacral region (60%), upper thoracic (10%) and cervical regions (5%). Because of the the rarity of this type of dermoid cyst, we would like to report this case of a 17 year old male who presented with right leg weakness and difficulty in walking. Keywords: Spinal dermoid, intramedullary, MRI, intraspinal tumour.

INTRODUCTION
Intraspinal dermoid cysts are rare and benign congenital tumors that occur due to the defective closure of the neural tube. They can be intramedullary (rare) & extramedullary (common). They comprise 1% of intracranial and intraspinal tumors [1]. Most common location is lumbosacral region (60%), upper thoracic (10%) and cervical regions (5%) [2]. Spinal dermoid can be uni or multilocular cystic tumors lined by squamous epithelium containing skin appendages (hair follicles, sweat glands, sebaceous glands) [3]. Spinal dermoid tumor can occur after myelomeningocele repair.

Inclusion cysts of the cord are rarely intramedullary, with only few isolated cases been reported. Only six cases have been reported in the literature [4].

CASE REPORT
A 17 year old male presented to the surgery department with the complaint of pain, difficulty in walking and decrease in power with reduced in sensation in the right lower limb since 3 months. These complaints were mild to moderate 3 months back and were on and off but have been aggravated in the last 3 months. The patient also complaints of lower back ache since 1 month. In general motor examination the Romberg’s test was found to be positive with power in right lower limb was 4/5 and left 5/5. After treating the patient symptomatically the following investigations were carried out. Blood profile and other serums were found to be of normal value. A detailed thoracic spine evaluation on MRI revealed A well-defined ablong intramedullary lesion noted in spinal cord extending from second to fifth thoracic vertebra measuring approx. 7 x 1 x 1.5cm (CC x AP x T) causing cord expansion at this level. The lesion shows both fat and poorly enhancing soft tissue component (Figure 1 A, B). The fat component appears hyper intense on both T1W & T2W (Figure 1 A,B) hypointensity on STIR MR images, findings consistent with fat (Fig 3). The fat component appears hyperintense on both T2W & T1W (Figure 2 A & C) while the soft tissue component appears heterogeneously hyperintense on T2W & heterogeneously hypointense on T1W (Fig 2 B & D). Spina bifida at 6th and 7th thoracic vertebrae is also noted. A CT scan of the same area revealed the same lesion with fat as its contents. The patient was then transferred to the neurosurgery department and taken for excision. A histopathological report confirmed the diagnosis of intramedullary dermoid.

Fig-1: (A, B) Sagittal T2WI & T1W showing well defined ablong intramedullary lesion in spinal cord extending from second to fifth thoracic vertebra fat component appears hyperintense on both T1W & T2W and soft tissue component heterogeneously hypertintense on T2WI and heterogeneously hypointense on T1W

Fig-2: (A-D) Axial T2WI & T1WI shows fat component as hyperintense on T2W &T1W and soft tissue component as heterogeneously hyperintense and heterogeneously hypointense on T1WI

Fig-3: STIR MR image fat component hypointense on STIR MR images

DISCUSSION
Dermoid cysts are rare benign lesions accounting for 1%–2% of all intraspinal tumors[1]. They occur between the 3rd and 5th weeks of embryonic life from the inclusion of ectodermal cells at the time of closure of the neural groove. Dermoid cysts develop during the embryonic period, they typically do not manifest until the 2nd or 3rd decade of life[5].

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They most commonly occur in lumbosacral region. Within the spine, an extramedullary location (60% of cases) is more common than an intramedullary location [5]. Spinal dermoid cysts are uncommon overall but account for nearly 20% of intradural tumors seen in patients younger than one year of age. They generally present in patients younger than 20 years[3].

Spinal dysraphism is commonly associated with dermoids. Associated congenital abnormalities include myelomeningocele, hypertrichosis, a dural sinus tract, or some combination of these abnormalities [5]. The same process which is responsible for the development of dermoid cysts gives rise to spinal dysraphisms. The incidence of dermoid in the cervical region is relatively low likely due to 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[4].

After myelomeningocele repair dermoid tumor may occur due to inadequate excision of dermal elements during dissection of placode or dura mater from the skin or with implantation of dermal elements inside the repair site. The growth of the entrapped skin may predispose the development of dermoid tumor[6].

Clinically intraspinal dermoids are important group of intraspinal tumors as they produce symptoms of spinal cord or root compression like other intraspinal space occupying lesion. Spinal dermoids usually cause gradually progressive symptoms of spinal cord or root compression before the diagnosis is established [5].

Choice for the work-up of suspected intraspinal cord lesions & current imaging modality is MR imaging. Traditional imaging work-up including radiographs, myelography, and computed tomography (CT) with and without intrathecal contrast material is replaced by MR imaging[5].

Diagnostic of dermoid tumour is combination of fat, fluid, soft tissue & calcium. MRI typically demonstrates heterogeneous signal intensity due to varying amounts of soft tissue, fat, calcium, and hemorrhage. The two major component of dermoid tumor are a lipid part and a more solid or more fluid part. Fat relatively have high signal on MRI especially the bright signal on T1W images, makes identification of the lipid component easy, because of increased water content the lesion can also appear hypointense on T1W images. This can be confirmed by demonstrating low attenuation (negative Hounsfield numbers) on a nonenhanced CT scan. The lipid component exhibit low signal intensity on gadolinium-enhanced fat-suppressed T1-weighted MR images. Fluid components are hyperintense on T2W images & occasionally May depict fat-fluid level. Cystic portion of the lesion does not typically show enhancement after the intravenous administration of contrast material; however. If soft-tissue component exists, it will usually enhance [1,5].

The treatment of choice for the dermoid tumor is the total excision of the mass at an early stage. Total mass excision is possible for extramedullary dermoid cysts; however, in intramedullary dermoid cysts, the capsule adheres to the cord and often leads to difficulties in their complete resection through surgery [7].

The common differential diagnosis of intramedullary cystic lesions is arachnoid cyst and epidermoid cyst. The arachnoid cyst shows cerebrospinal fluid (CSF) signal with no diffusion restriction whereas epidermoid shows hyperintense signal in fluid attenuation recovery sequence and restriction of diffusion [5].

CONCLUSION

Within the spine, an extramedullary location (60% of cases) is more common than an intramedullary location. Choice for the work-up of suspected intraspinal cord lesions & current imaging modality is MR imaging

REFERENCES