

Spinal Intramedullary Dermoids: Report of 2 Cases and Review of Literature

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ABSTRACT

Dermoids are rare congenital inclusion tumours of spinal canal with a reported incidence of less than 1% of all spinal neoplasms. They are commonly found in intradural location and are very rare in intramedullary compartment. Very few cases have been reported so far in literature. We report here two cases of dermoids in intradural and intramedullary location in view of the rarity of the lesions. We discuss the clinical features, imaging characters and microsurgical excision techniques along with review of the relevant literature.

Keywords: Dermoid cyst, spinal cord tumour, magnetic resonance imaging.

INTRODUCTION

Dermoids and epidermoids are rare congenital developmental cysts found anywhere along the entire neuroaxis, thought to be a consequence of embryological errors during neural tube closure. Though rare they have to be considered in the differential diagnosis of other spinal cord tumours and a high index of suspicion is necessary particularly when they present with dysraphic anomalies of the spine. Since, they are benign slow growing lesions, total excision of the cyst with microsurgical techniques where ever possible offers total cure without the possibility of recurrence.

CASE REPORTS

CASE 1

7 year old male child presented with difficulty in walking for the past 1 year. It was a progressive weakness of both lower limbs started insidiously and is able to walk with

support only now. He developed urinary incontinence and constipation in last 2 months. There was no history of trauma.

General examination was apparently normal and he had spastic paraparesis of 2-3/5 power in both lower limbs with brisk DTRS and absent superficial reflexes. Both plantars were upgoing. Sensory level was at D6-7. His routine blood counts and biochemical analysis was normal. X-ray dorsal spine did not show any gross abnormality. MRI spine showed a large hypointense mass lesion in T1wi opposite C7-D4 with expansion of the cord [Figure 1:a&b]. There was an extramedullary component also noted. The tumour had hypointense cystic areas at places. On T2wi the lesion showed hyperintensity with a small tract going in between the lamina up to the surface. There was no contrast MRI film but a diagnosis of dermoid was kept in mind apart from other intramedullary pathologies like astrocytoma and ependymomas as differential diagnosis.

He was operated by D1-D5 laminectomy. On opening the dura there was a large well defined lesion containing soft putty and pultaceous material with lots of hair in side. The lesion along with the tract excised completely under high power magnification using sharp microdissection. Most of the capsule could be easily peeled off as it was very thin but at few areas it was adherent due to fibrosis hence no attempt was made to separate it from the cord. Dura closed in water tight fashion after excising the dermal tract.

Post-operatively he made an excellent recovery by two weeks period. Histopathological examination of the cyst showed lining by stratified squamous epithelium surrounded by fibrocollagenous tissue and sweat glands in the stroma [Figure 1:c].

CASE-2

3 year old male child was brought with complaints of difficulty in walking for the past two weeks and history of fever since one week. There was h/o bowel, bladder incontinence also for the past two weeks.

On examination, child had a midline dimple which was present since birth at the lumbosacral region. Neurological examination showed spastic paraparesis with 3/5 power in both lower limbs and brisk DTRs. Sensory examination was not possible.

His blood counts were normal. X-ray Lumbosacral spine was normal and there was no evidence of spina bifida. MRI spine showed a large well defined mass lesion opposite L1-4 region hypointense on T1WI and hyperintense on T2WI with predominant cystic areas opposite L2 and L3 vertebra. There was expansion of conus and filum was seen low down attached to S2 [Figure 2: a&b].

Child was operated in prone position by L1-5 laminectomy. There was a subcutaneous tract from the midline dimple going intradurally in to the mass. The mass contained pearly white flakes of keratin material and some hair also inside [Figure 2: c & d]. The lesion was extending in to conus pushing all the roots to one side. The cyst had a very thin layer of capsule which was gently separable from the roots and from the conus. De-tethering of the cord done by dividing the filum at the lower end and dura closed in water tight fashion after thorough saline irrigation of the tumour bed. Post-operatively he recovered well.

There was CSF leak, for few days with small wound gape but ultimately settled in two weeks time and by his last follow-up he is active and self ambulant and regained bladder control in 3m time. Histopathological examination of the tumour was consistent with dermoid [Figure 2:e].

DISCUSSION

Dermoids are rare congenital inclusion cysts of spinal canal with a reported incidence of <1 %. They are common in children accounting for 5-17% of all intradural spinal lesions in reported series. They are often found in association with spinal dysraphic abnormalities and are intradural in location.

Intra medullary location is very rare and only few case series have been reported so far.^[1] They are thought to arise from ectopic foci of dermal appendages retained during the 3rd-5th week of embryonic life before neural tube closure. It is postulated that the timing of the event, early or late determines the cell potentiality which in turn determines the type of the tumour (epidermoid, dermoid or teratoma). They are common at the poles as this portion of neural tube closes at a later period.^[2] Like epidermoids they have a thin walled capsule lined by stratified squamous epithelium and contain derivatives of dermal appendages.

They grow by accumulation of keratin and active secretion from dermal appendages and sweat glands. They often have a communication to the exterior by means of a tract which leads to repeated attacks of infections.^[3] But patients presenting with recurrent meningitis itself is very rare.

The clinical presentation of spinal dermoids is by compressive pathology similar to other space-occupying pathologies in the spinal canal. Spastic or flaccid weakness of limbs coupled with bladder disturbances are the common presenting findings. Often there may be associated evidence of spinal dysraphism, like a dermal sinus tract in the midline may be found especially in children

Imaging findings are almost certain for epidermoids and dermoids. The characteristic findings include heterogeneous signal intensities in a well defined lesion in the intradural/intramedullary plane, reflecting the contents of the cyst. T1-weighted images show hypo to isointense well defined lesion with hyperintense foci if the cyst contain more fat and triglycerides and hyperintense on T2-weighted images.^[3]

Satellite lesions at adjacent levels are not uncommon. They generally do not enhance following contrast injection but in some cases thin peripheral enhancement noted suggestive of a matured cyst lining or of chronic fibrotic reaction surrounding the capsule due to inflammatory response. Also careful examination may give clue regarding the presence of subtle dysraphic abnormalities of spine if present.

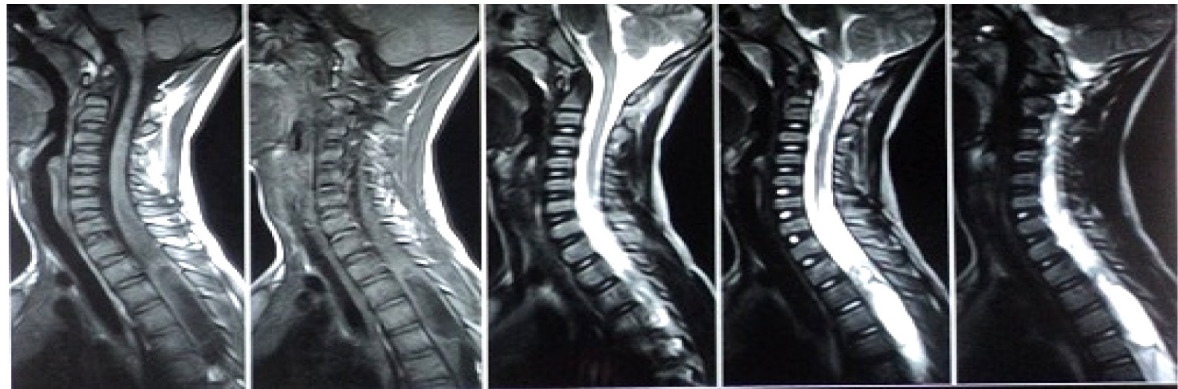


Figure-1 a) MRI Spine T1WI-showing large well defined hypo intense oval shaped mass lesion in intradural plane opposite D1-D4
b) T2 WI-axial images of dorsal spine showing hyperintense signals in the mass compressing and pushing the cord to one side

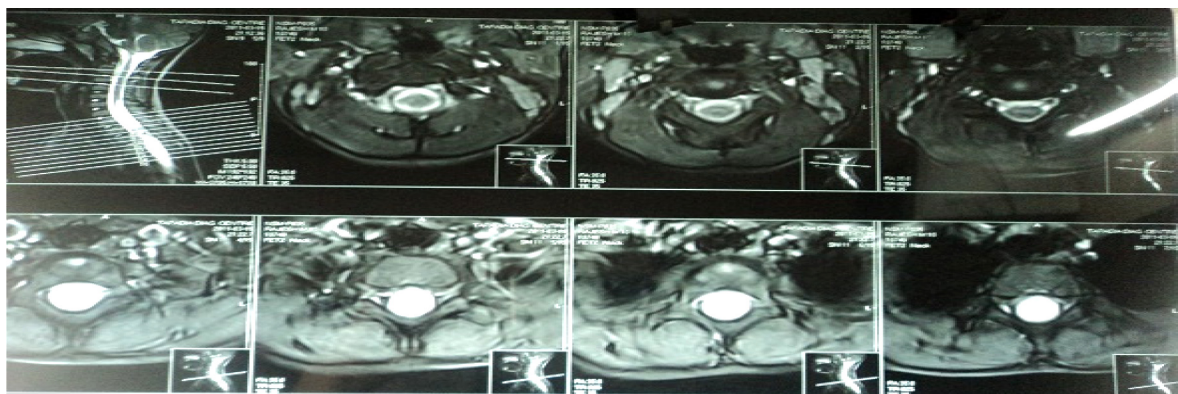


Figure-2 a) Large well defined irregular mass lesion iso intense with hypointense cystic spaces in between seen in intradural plane opposite L3 ot-S1,2 space
b) MRI-axial images showing heterogenous signals in the same mass
c) & d) Intra-op photographs showing the tumour with white keratin flakes and hair inside the cyst.

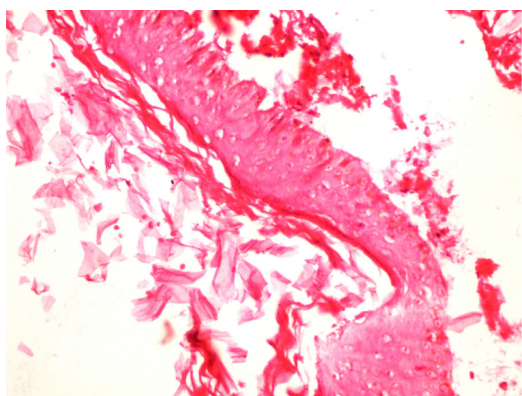


Figure 2 : c) Histology of the excised specimen

The management protocol include total to maximum resection of the lesion including the sinus tract if present in symptomatic individuals. Total resection of the capsule may not be possible always in intramedullary plane. [4, 5] In such cases maximum resection can be tried with microsurgical techniques without endangering the neural elements and small bits of capsule at adherent areas can be left over.

Patient will have good symptomatic relief and as these are benign slow growing lesions they can be followed up at regular intervals as long as patient have a symptom free life. [6] Asymptomatic tumours found incidentally on routine imaging at critical areas can be

followed up and are operated as and when patient become symptomatic.

CONCLUSION

Intraspinal dermoids are rare benign congenital inclusion tumours of spinal canal and are to be considered in differential diagnosis of intradural neoplasms of spinal cord with heterogenous signal intensities, especially in children presenting with spinal dysraphic abnormalities like dermal sinus tracts. Careful MRI screening of the spine may reveal subtle dysraphic anomalies and helps in preoperative diagnosis with certain, even in cases who doesn't show external evidence of midline dermal sinus. Microsurgical total excision of the lesion wherever possible offers best prospects of cure without recurrence.

CONFLICT OF INTERESTS

We declare that we have no conflicts of interest.

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