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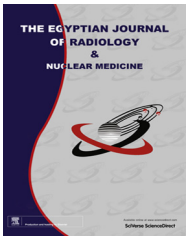

CASE REPORT

Ruptured thoracic intraspinal dermoid cyst in a patient with skeletal abnormalities of thoracic spine – A case report

Mrudang Raval [a,*], **Sunita Purohit** [b], **Vineet Mishra** [c], **Tanay Shah** [a]

[a] *SDMH, Jaipur, India*
[b] *Radiodiagnosis Department, SDMH, Jaipur, India*
[c] *Vardhman Scanners and Imagers, SDMH, Jaipur, India*

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KEYWORDS

Spinal dermoid;
Diastematomyelia;
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Magnetic resonance imaging;
Hemi vertebrae

Abstract Spinal dermoids are congenital multi- or unilocular benign cystic tumors lined by squamous epithelium containing skin appendages. The tumours become acutely symptomatic following infection or rupture. We present a very rare case of ruptured thoracic spinal dermoid cyst in a 12 year old girl with congenital vertebral abnormalities, who presented with back ache of recent onset and a 5 year history of unnoticed slipping of footwear while walking. Magnetic resonance imaging reveals a lesion in the thoracic spine with fat droplets in bilateral frontal horns of lateral ventricle and cisterns of brain. Other additional findings were also noted.

1. Introduction

Dermoid cysts are rare benign lesions within the central nervous system that were first described by Verratus in 1745 and further popularized as the "pearly tumor" by Cruveilhier in 1829 (1,2). The etiology of dermoid cysts is not completely understood, and theories for both congenital and acquired etiologies exist (2–4). The lumbosacral region is the most common site to be affected (60%) (5). They are benign slow growing lesions which become clinically apparent during the second or third decade of life (6) and males are more commonly affected. We present a very rare case of a ruptured thoracic spinal dermoid cyst with fat droplets in both lateral ventricles and cisternal spaces of brain in a 12 year old girl with congenital vertebral abnormalities.

2. Case report

A 12 year old female presented with a 4 day history of back pain. Pain was mild dull aching without any radiation. Patient had a chronic 5 year complain of unnoticed slipping of footwear. Developmental mile stones were properly achieved without any delay. On neurological examination no abnormality was detected. Gait was normal.

X-ray thoracic spine anteroposterior view and lateral view was done. X-ray showed an exaggerated kyphosis of thoracic spine with butterfly and hemivertebrae from D6–D9 levels.

* Corresponding author.

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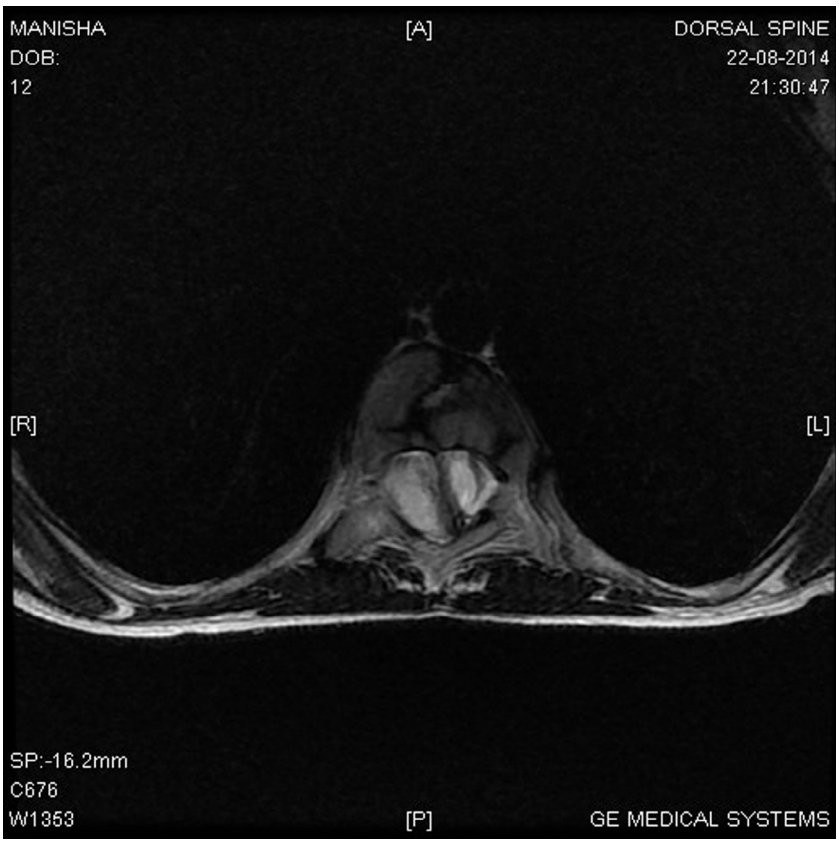

Fig. 1 T2w axial sections at D8–9 level showing a bony spur dividing the cord into two hemicords.

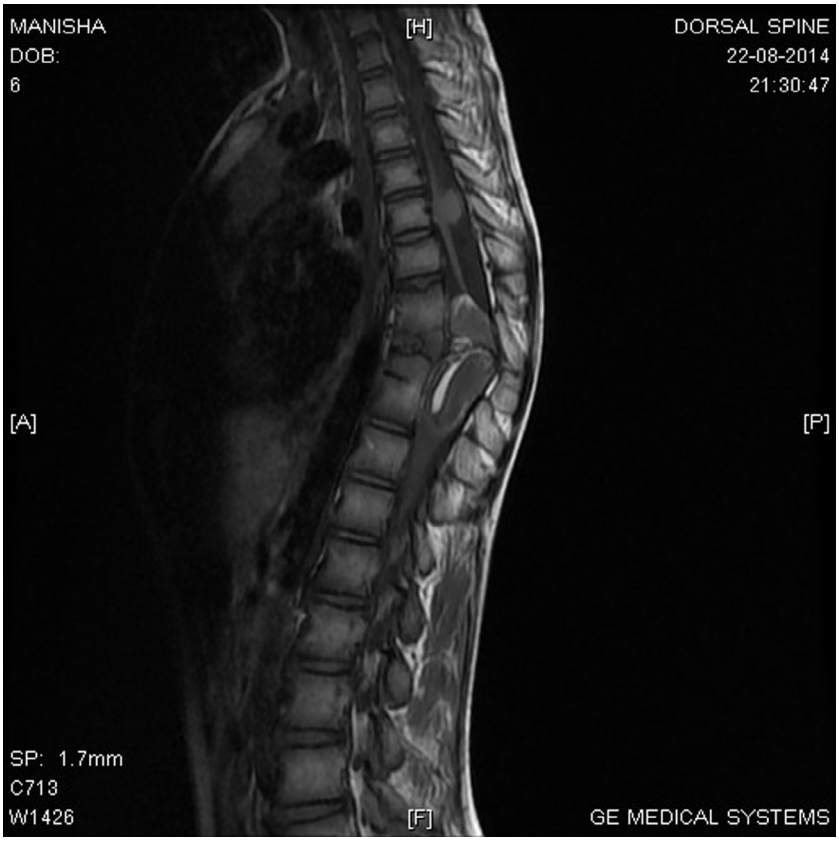

Fig. 2 Sagittal T1w section of brain reveals scattered hyperintensities in ventricles and cisterns which suppressed on fat saturation technique suggestive of fat droplets from ruptured dermoid.

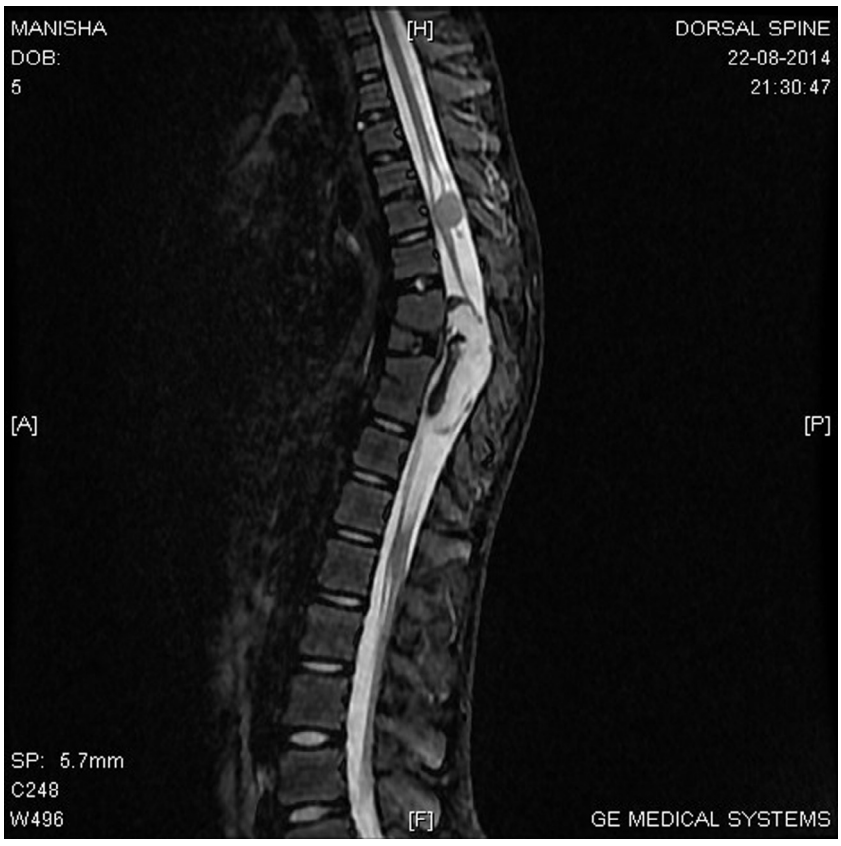

Fig. 3 STIR sagittal image of thoracic spine showing nodular lesion at D4 level which is hypointense and oval lesion at D6–10 level which is hyperintense except for the fat component.

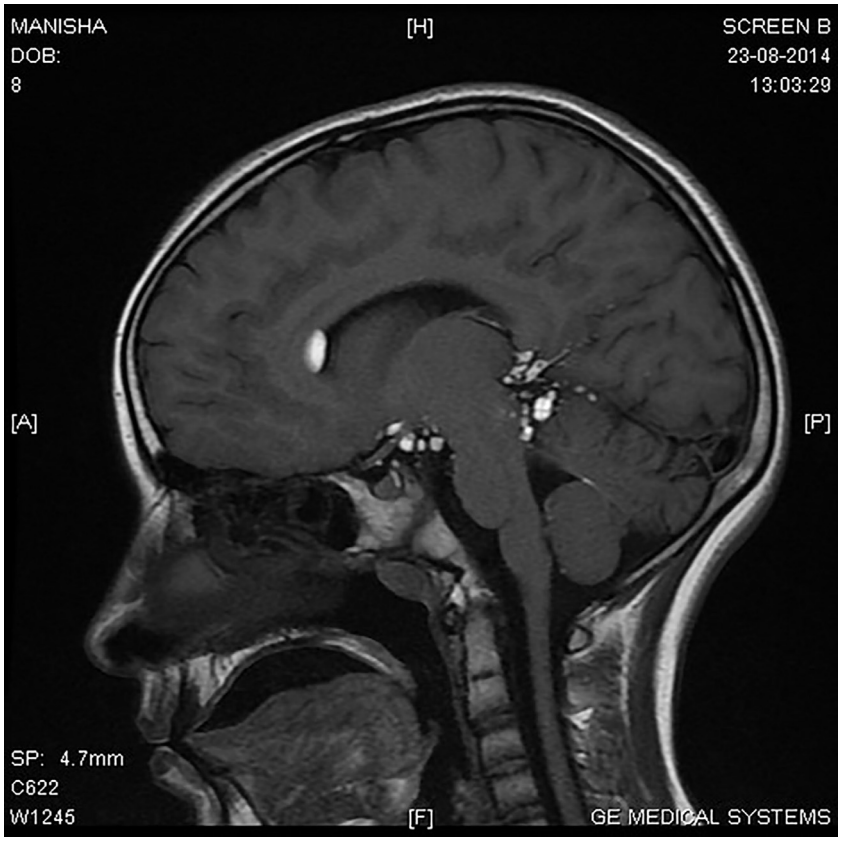

Fig. 4 T1w sagittal image of thoracic spine showing a nodular mass lesion at D4 level and an oval mass lesion at midthoracic level showing hyperintense fat component within.

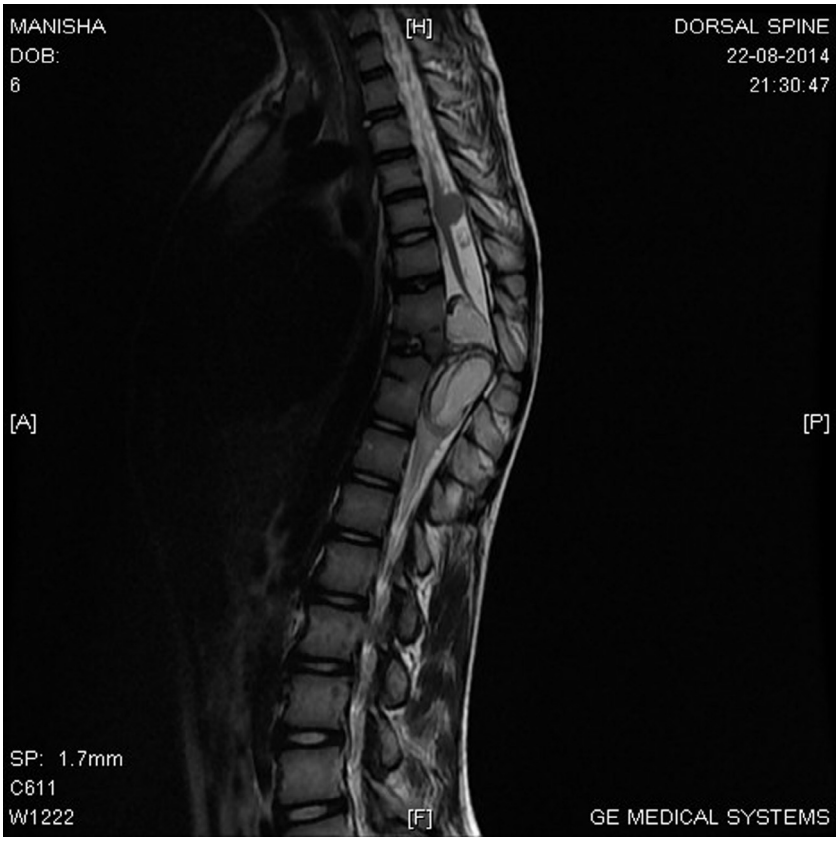

Fig. 5 T2w sagittal image of thoracic spine showing hypointense neurofibroma at D4 level and hyperintense oval mass lesion within isointense fat component.

A horizontal bony ridge was seen at the level of D8 vertebra extending into the spinal canal which probably raised suspicion of diastematomyelia. Rest of the thoracic spine and disk spaces were normal.

Magnetic resonance imaging was performed on 1.5 T GE-HDXT with fat suppression studies and without use of intravenous contrast material. T1- and T2-weighted spin-echo and short inversion time inversion-recovery (STIR) MR images were acquired in the axial, sagittal and coronal planes. T1w and Fluid attenuation And Inversion Recovery (FLAIR) Images of brain were obtained. Study revealed a well defined $51 \times 19 \times 20$ mm intramedullary mass lesion extending from D6 to D10 vertebrae. No evidence of spinal dysraphism was noted. The major part of the mass was hyperintense on T2w images and hypointense on T1w images suggesting fluid component within the lesion. A part of the mass was hyperintense relative to CSF on T1w and isointense to CSF on T2w images and hypointense on STIR consistent with fat. Thinning of the cord parenchyma was noted surrounding the lesion. Skeletal abnormalities of spine in the form of butterfly vertebrae, hemivertebrae and block vertebrae were noted on 6th, 7th, 8th and 9th thoracic vertebrae (see Figs. 1–3).

A hypointense bony ridge was noted, dividing the spinal cord into two hemicords at the level of D8–9 vertebrae suggesting diastematomyelia. A well defined extramedullary intradural nodular lesion measuring $13 \times 13 \times 11$ mm hypointense on T1, T2w, and STIR images on the dorsal aspect of the cord, compressing the cord ventrally, at the level of D4 vertebra was noted as an additional finding suggestive of a neurofibroma (see Figs. 3 and 5).

FLAIR and T1w axial and sagittal sections of brain revealed multiple fat droplets in frontal and temporal horns of right lateral ventricle infra- and supratentorial cisterns and few sulcal spaces which were hyperintense on FLAIR and T1w images and suppressed on fat saturated sequences (see Fig. 4).

Provisional preoperative diagnosis of ruptured dermoid cyst with fat droplets in lateral ventricles and cisternal spaces of brain was given. Additional findings included vertebral segmentation anomalies, diastematomyelia and neurofibroma (Fig. 5).

Patient was operated with laminectomy and near total excision of the lesion. Intraoperatively, the lesion was intramedullary, white cheesy with hair inside.

Histopathological examination showed a cyst wall lined by stratified squamous epithelium with the presence of sebaceous glands and hair follicles inside. In view of the presence of hair on gross examination the diagnosis of dermoid cyst was given.

3. Discussion

Diastematomyelia is a rare congenital malformation of the spinal cord, which belongs to the group of occult spinal dysraphisms (7). It was first described in 1837 by French researcher C.P. Ollivier, who used the term ‘diastematomyelia’ (8). A new classification and term (split cord malformation, SCM) was introduced by Pang (9), who distinguished two types of diastematomyelia. In type I (SCM type I), two parts of the split cord (hemicords) are placed in their separate dural

tubes. The spinal canal is divided into two by a septum, osseocartilaginous in most of the cases (75%) or, rarely, a fibrous one (25%). Type II (SCM type II) is characterized by the presence of one common dural tube, without any osseous septum in most of the cases, or with a fibrous septum, if any. Type II cases are asymptomatic (7–11).

Diastematomyelia is more common in women (80–90%) and tends to appear within the thoracolumbar spine (85%). It is rarely confined to the lumbar or to the thoracic spine solely. There were also cases of cervical or multilevel diastematomyelia (7,10,11). Frequently coexisting skin lesions include capillary angiomas (26%), dermal sinuses (22%), subdermal lipomas (11%) or other dermal naevi and crypts (12–16). Moreover, the diastematomyelia may be accompanied by spinal abnormalities such as hemivertebrae, butterfly vertebrae, fusion of vertebrae and narrowing of the intravertebral space. Very often there is also a scoliosis – even in 30–60% of cases (12,14–16). There were also cases of tethering of the spinal cord or of the terminal cone solely, syringomyelia, Klippel-Feil's syndrome, epidermoid and dermoid cysts, lipomas and dural cysts, especially the arachnoid cysts. Diastematomyelia is present in 15–20% of cases of Arnold-Chiari II syndrome (14,15).

Dermoid tumors are rare, benign, congenital lesions, which comprise 1% of intracranial and intraspinal tumors (17). Dermoid tumors are typically unilocular cysts containing yellow or yellowish-brown viscous fluid with creamy contents of different types of fat (crystals of cholesterol, lipid metabolites, and keratin). High lipid content emanated from sebaceous glands causes high signal intensity on T1W spin echo images (18). The signal intensity may be heterogeneous related to different components in the cyst (19). Bone and cartilage might be found in the tumor itself and sometimes calcification can be seen in their walls.

Spinal dermoid tumors can be intramedullary, intradural-extramedullary, or extradural (11,20). They occur predominantly in the lumbosacral region (60%), involving the cauda equina and conus medullaris, and are quite rare in the upper thoracic (10%) and cervical regions (5%) (6). A dermoid cyst can rupture during surgery, after a trauma, or spontaneously (21). If rupture has occurred, contents of the cyst will spread out along the subarachnoidal space and ventricular system. As was noted in our case.

Neurofibromatosis in our case was an additional finding. No such association between neurofibromatosis and diastematomyelia was found.

4. Conclusion

Diastematomyelia is a rare dysraphic defect of spinal cord and is associated with various anomalies and abnormalities of spine. Spinal dermoids are associated with diastematomyelia. Ruptured dermoid cyst can lead to dissemination of fat droplets along the CSF pathway into the subarachnoid space and basal cisterns of brain.

Conflict of interest

There is no conflict of interest.

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