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Case Report

Type II diastematomyelia with intramedullary epidermoid cyst: An expected but rare association

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Abstract

Diastematomyelia is rare congenital anomaly most often seen in lumbar region .Epidermoid tumors in spinal canal are rarer, more so in the intramedullary location. While the pathogenesis of diastematomyelia has been explained in classical papers and can explain the co-occurrence of epidermoid tumors in these cases, the actual reported cases as such are quite rare. The author reports clinical, radiological and operative findings of such a case.

Keywords: Spinal cord tumour, Spinal dysraphism, Epidermoid tumour, Lumbar

1. Introduction

Diastematomyelia is rare congenital anomaly. The embryological defects leading to it have been previously explained in the classical paper by Pang *et al*.[1] Rare reports of association of diastematomyelia with various kinds of epithelial inclusion cysts and other tumors have been appearing in literature[2,3]. This co-occurrence of inclusion cysts and other tumors is expected on the basis of the unified theory of split cord malformations[1] but few such reports are available in literature. Such a case of thoraco-lumbar diastematomyelia associated with intra-medullary epidermoid cyst of spinal cord is reported here.

2. Case Study

A three year old girl presented to us with subacute onset weakness of both lower limbs occurring over a period of four weeks. This was associated with urinary incontinence and severe pain in both lower limbs which made it difficult of her to sleep. On examination, she had presence of an abnormal tuft of hair over thoraco-lumbar junction region and bifid spinous processes were palpable underlying it. Examination of the lower limbs revealed spastic paraplegia. However, there was no cutaneous sensory disturbance barring paraethesiae. Ultrasonography of the abdomen revealed no other abnormality. CT of thoraco-lumbar region revealed bony midline septum at L2 level dividing the spinal canal into two. MRI of the region of interest revealed two separate dural sacs at the same level which were united both proximally and distally. In addition, it revealed presence of a cystic lesion in the cord at D11-D12 levels which was hypointense on T1, hyperintense on T2 (Fig 2), with contrast enhancement at the lower pole (Fig 1).

Fig 1. Intramedullary lesion with maximal enhancement at lower pole.

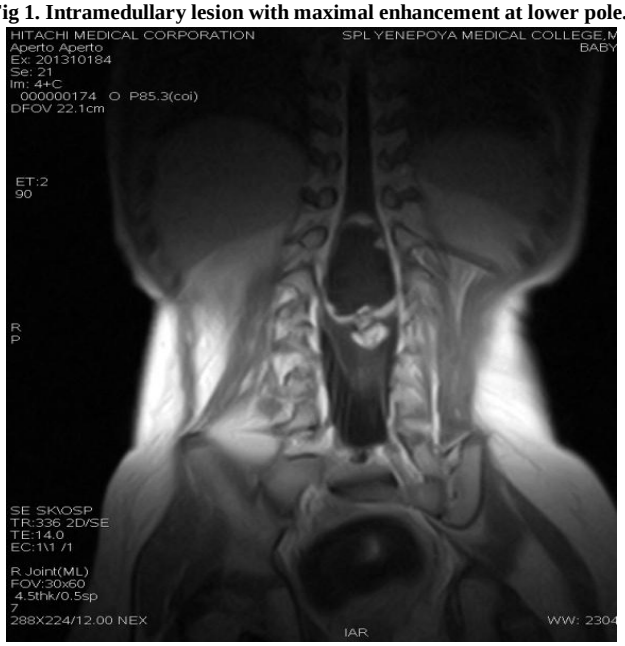
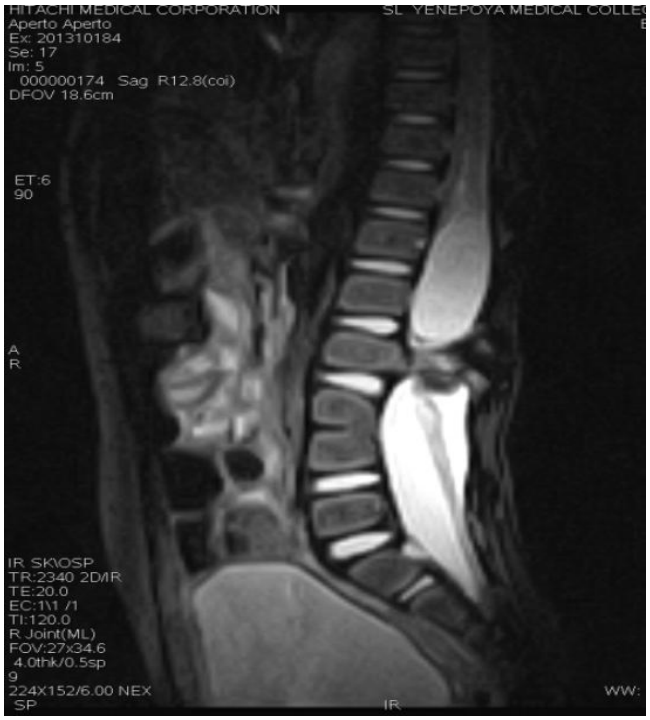


Fig 2: T2 weighed MRI showing the cystic lesion to be hyperintense. Hypointense sagittal bony septum is also seen along with L2-L3 block vertebra. Taut and thick filum terminale is evident.



3. Results

Excision of the bony septum and epidermoid tumour was performed. Adhesions between the thecal sac and cord were also lysed. Post-operative period was uneventful. Histopathological examination of the cyst wall revealed stratified squamous epithelium. The child made dramatic improvement and was ambulant independently at sixteen weeks with some residual spasticity.

4. Discussion

Diastematomyelia has been classified into type I and II depending on whether the split cord resides in a single or separate dural sacs. Accordingly, the present case is a type II diastematomyelia. As per unified theory of Pang *et al*[1], the error is attributed to appearance of an accessory neurenteric canal through the embryonic disc that maintains communication between yolk sac and amnion and thus enables contact between the ectoderm and endoderm within the canal.

Intramedullary epidermoid tumors of spinal cord arise as a result of displacement of ectodermally committed cells at the time of neural tube closure around third to fifth weeks of development. Occasionally, they arise due to traumatic implantation at lumbar puncture.

Few reports of diastematomyelia and epidermoids are available. The present case provides further support to the unified theory of embryogenesis for double spinal cord malformations.

References

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