CASE REPORT

THE RARE ASSOCIATION OF DIASTEMATOMYELIA WITH LIPOMYELOCELE AND DORSAL DERMAL SINUS - CASE REPORT AND LITERATUE REVIEW

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ABSTRACT

We report the MRI dorsolumbar spine of 10-year child who was referred to the Civil Hospital in June 2016 with symptoms of backache for last one month and birth history of congenital discharging sinus. Magnetic Resonance Imaging of his spine revealed type A diastematomyelia, multilevel vertebral segmentation anomalies, anterior and posterior spina bifida and tethered cord,along with dorsal dermal sinus and lipomyelocele. Magnetic Resonance Imaging (MRI) is the investigation of choice for diagnosing occult spinal dysraphisms.[1] We present the rare case of triple occult spinal dysraphic anomaly of different embryological origin in a single patient.

Keywords: diastematomyelia; lipomyelocele; MRI; tethered cord; dorsal dermal sinus, occult spinal dysraphism

Introduction

Ocultspinal dysraphism (OSD) is characterized by failure of fusion of the vertebral arches, with unexposed neural tissue; the skin overlying the defect is intact.[2] The spectrum of OSD comprise lipomyelocele, lipomyelomeningocele, meningocele, myelocystocele, filar and intradural lipomas, persistent terminal ventricle, dermal sinuses, complete dorsal enteric fistula to neurenteric cysts, diastematomyelia and caudal agenesis. MRI is the imaging modality of choice for evaluation of this complex group of disorders. Diastematomyelia is a rare form of occult spinal dysraphism, occurs secondary to splitting of the notochord. It is defined as sagittal division of the spinal cord into two hemicords. There can be two dural sac with a fibrous band or bony spur within the cleft (type A) or a single dural sac surrounding them (type B). Type A is associated with segmental anomalies of the vertebral bodies. Clinically, it might present with dermatologic finding or tethered cord syndrome.[3-5] Radiological diagnosis for tethered cord includes filum terminale thickness greater than 2 mm or conus medullaris position below L1-L2 disc level on MRI.[6] On the other hand, Lipomyeloceles and lipomyelomeningoceles that represent another spectrum of OSD, occurs due to interposition of mesenchymal tissue between the surface and neuroectoderm; are characterized clinically by the presence of a subcutaneous fatty mass above the intergluteal crease. The main differentiating feature between the two is the position of the placode-lipoma interface, lying within or outside of the spinal canal respectively. Dorsal dermal sinus- an epithelial lined fistula that connects neural tissue or meninges to the skin represents another category of OSD due to incomplete separation of the superficial ectoderm from the neural ectoderm.[7] We present a rare case of type A diastematomyelia associated with lipomyelocele and dorsal dermal sinus in a 10 year old child.

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Case Summary

A 10 year old boy was referred to the Civil Hospital Karachi, with complaint of nonspecific lumbalgia along with lump over his back since birth. There was past history of discharging sinus in the back at birth. He had no motor or sensory problems, nor has he had any bowel or bladder issues. Developmental and family history was not significant. Blood chemistry and urine analysis were normal. The physical exam including the neurological examination was unremarkable except for tender 8x5 cm soft mass above the natal cleft at midline with the central non discharging pit. Neuro-Imaging of the Dorsolumbar spine was performed on GE Health Care Signa HDxt 1.5 Tesla MRI Scanner, using the four-element phased-array body coil with slice thickness of 4 mm with interslice width of 1 mm. Multiplanar, multisequential images in T1W, T2W fast spin echo sequence, STIR along with fat suppressed post contrast T1W sequence were obtained. MRI revealed multiple anomalous vertebra seen varying from hemi-vertebra, block vertebra and butterfly vertebra involving the distal dorsal and lumbar spine between D10-L4 vertebrae. There is non fusion of the neural arches of the involved vertebra with widening of the interpedicular distance. No evidence of radicular or nerve root compression was seen at any level. There is sagittal splitting of the spinal cord into two hemicords at D12 vertebra which reunite at L4 vertebra with intervening fibrous septum. Each hemicord is contained in its own dural sac. Low conus medullaris below L2 reaching upto L4 level. No hydromyelia was seen. It was associated with non enhancing high T1, T2 and low STIR signal mass in the epidural space invading the subarachnoid space extending from D10-L4 level. Posteriorly, it is merging with the subcutaneous (similar signal intensity) mass on back. It measures 12.4.3x5.8x5.1 cm in CCxTVxAP dimensions. It is causing widening of the spinal canal, splaying of the posterior elements of the spine. There is linear hypointense T1 and T2 tract extending form skin at the level of L2 vertebra for the length of 4.1 cm in the above mentioned lesion.

Discussion

Our case describes a large epidural-subcutaneous lipomatous mass on MRI in a 10 year old child. The lipoma-neural placode interface was seen within spinal canal, characteristic for lipomyelocele.[7] There was tethered cord and type A Diastematomyelia was identified associated with anterior and posterior multilevel dorsolumbar spina bifida. Hoffman et al[8] in the study of 97 patients with lipomyelomeningocele, reported anassociation with dermal sinuses and diastematomyelia in only 3.1% of the patients. In a cross-sectional study of Saima Sadiq et al,[9] conducted at the SIUT, Karachi, out of 110 patients with spina bifida, only 4 had diastematomyelia, none had lipomyelocele. In the case report of Lívia Teresa etal,[10] an

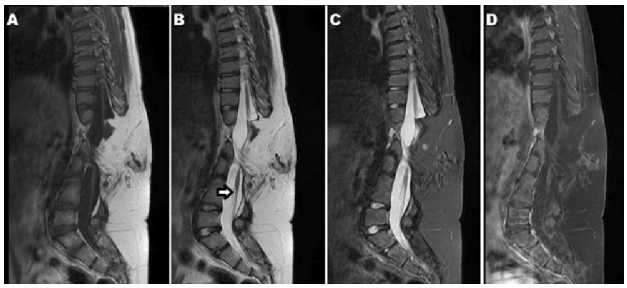

Figure 1: Mid-Sagittal T1-weighted image **(A)** and T2-weighted image **(B)**, STIR **(C)** and contrast enhanced fat suppressed T1-weighted image **(D)** of spine showing widening of spinal canal, tethered cord (arrow) and Lipomyelocele.

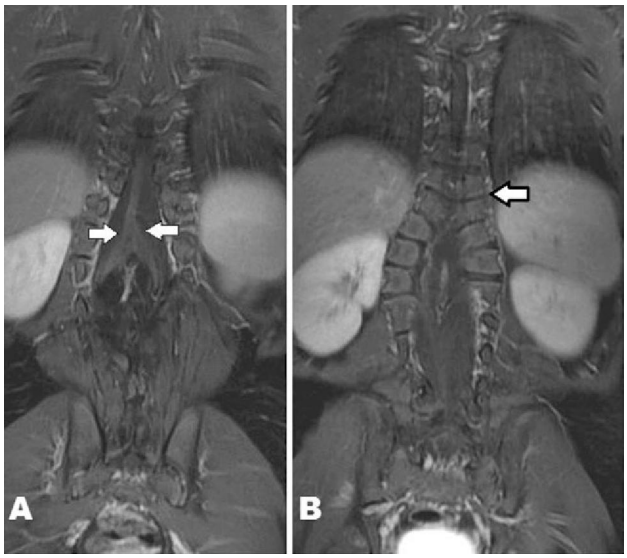

Figure 2: Coronal contiguous fat sat post contrast T1-weighted images **(A)** and **(B)** of spine showing diastematomyelia (arrow) and segmentation vertebral anomalies involving the dorso-lumbar spine.

association of spinal lipoma with congenital dermal sinus was described. In 2003, Guirish A. Solanki and his colleagues[11] described first case report of quadruple spinal dysraphic anomalies in a single child i.e:

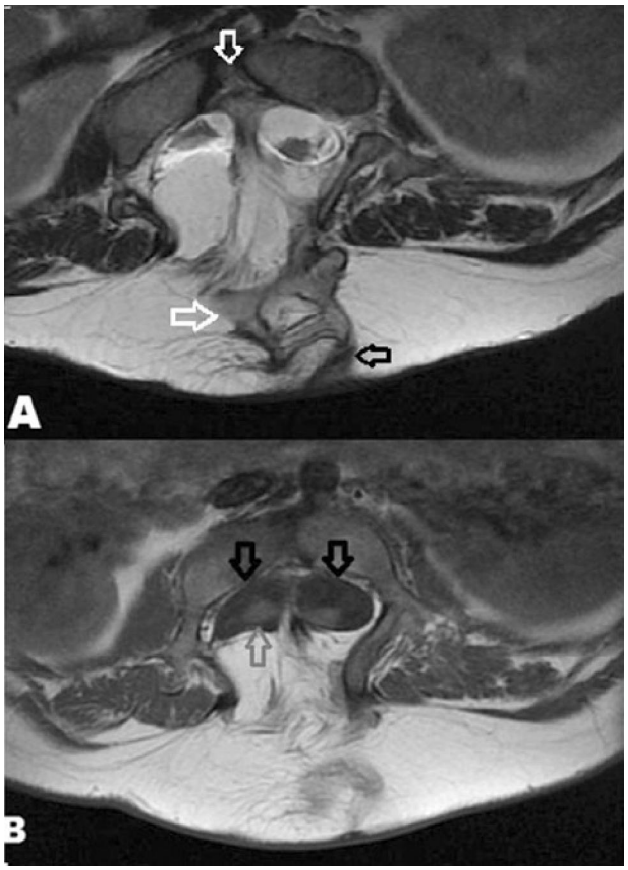

Figure 3: Axial T2-weighted image and T1-weighted image **(A)** and **(B)** of spine at level of L1 vertebra showing dermal sinus (black arrow in A), anterior and posterior spina bifida (white arrow in A), two dural sacs (black arrowsin Fig B) and lipoma neural placode interface seen within spinal canal (Gray arrow in Fig B)

meningocele, concurrent a type-1 split cord malformation (SCM) associated with hemivertebrae, lipomyelomeningoceles and terminal myelocystocele-comparable to our case. The triple occult coexistent spinal dysraphisms of different embryological origin in a single patient with this combination was unique to our case. This case highlights the importance of multiple coexistent spinal dysraphisms. Each deserves special attention, as each anomaly can lead to different complications. For example dorsaldermal sinus can lead to meningitis if patent and communicating with spinal canal and split-cord malformations are usually associated with tethered cord. Surgical intervention is almost inevitable because symptoms progress over time.The affected individual need psychosocial guidance and counseling.[12] Fortunately in our patient, no bladder, bowel, sensory-motor symptoms were seen, so followup visits were recommended.

Conclusion

The aim of this case study was to re-emphasize the role of Magnetic Resonance Imaging (MRI) in the diagnosis of multiple coexistent closed spinal dysraphisms and to document rare association of diastematomyelia with concurrent lipomyelocele and congenital dorsal dermal sinus.

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