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Research Article

AN UNUSUAL CASE OF MULTI-LEVEL SPINAL DYSRAPHISM: TWO UPPER THORACIC MENINGOCELES AND A LUMBAR SPINA BIFIDA OCCULTA WITH NO NEUROLOGICAL DEFICIT

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ABSTRACT
Spina bifida is a common congenital anomaly consisting of a wide spectrum of neural tube defects. We however, came across a rare presentation of this common entity and report here an unusual case of a child with concomitant two different levels of spina bifida cystica in the thoracic spine and one spina bifida occulta in the lumbar spine. This multi-level spinal dysraphism was not associated with any form of neurological deficit or other complications and the child appeared well on presentation.

Keywords: *Spinal dysraphism, Spina bifida, Neural tube defect*

INTRODUCTION

Neural tube defects result from failure of the neural tube to close spontaneously between the 3rd and 4th week of in-utero development (Kinsman et al., 2016). Spina bifida literally means "spine in two parts" or "open spine" (HarwoodNash *et al.,* 1991). Spinal dysraphism involves failure of bone fusion in the midline of the vertebral column resulting in a defective neural arch through which meninges or neural elements herniate, leading to various clinical manifestations. It is broadly classified as spina bifida aperta and spina bifida occulta (HarwoodNash *et al.,* 1991, Venkataramana *et al.,* 2011). In occulta, there is midline defect of the vertebral bodies without protrusion of the spinal cord or meninges and with normal skin cover (Kinsman *et al.,* 2016). In spina bifida aperta, the herniation is called meningocele or myelomeningocele, depending on whether the meninges herniate alone or along with spinal cord parenchyma and nerve roots. They are usually associated with skin defect (open defects) with an imminent risk of CSF leak (Sarnat *et al.,* 2006). Spina bifida has a multifactorial causation, encompassing both genetic and environmental factors. Recent information has stressed upon the importance of maternal nutrition and folic acid supplementation in its causation, which has contributed to the reduction in incidence of neural tube defects (Morrow *et al.,* 1998 & Pal-de Bruin *et al.,* 2000). The authors report here a rare case of multi-level spinal dysraphism in a neonate without any neurological deficit.

CASE

A 7-day old male child presented to the OPD with two discrete swellings in the upper back and a tuft of hair in the lower back since birth. There was no history suggestive of any focal neurological deficit in the form of decreased movements of limbs or urinary complaints. The parents did not give any history of discharge or leak from the swelling or fever. The child was otherwise active, feeding adequately and appeared well. He was the first child born of a non-consanguinous marriage by normal vaginal delivery at full-term with a birth weight of 3 kg with an uneventful antenatal and natal course. Hematinics were taken in the second trimester in the antenatal period and ultrasonography done at 8th month showed neural tube defect in the fetus.

On physical examination, we found two discrete swellings in the upper thoracic region of the spine, measuring about 6×4 cm and 4×3 cm respectively, soft in consistency, fluctuant, pedunculated and covered with partially epithelialized skin. The upper swelling was transilluminant while the lower was not. Along with this there was a tuft of hair in the lumbar region with no visible swelling. Muscle tone,

power and response to pain were normal in all the four lower limbs. Deep tendon reflexes were normal. Anal tone and reflex was also normal and there was no rectal prolapse or urinary dribbling. No bony deformity was present in the lower extremities.

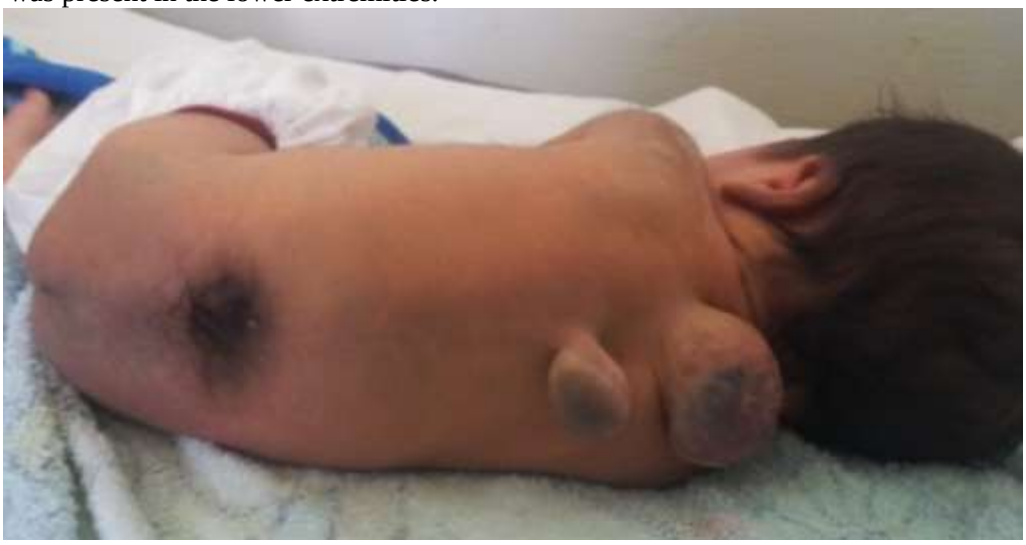

Figure 1: Spinal dysraphism at three levels.

MRI of the spine revealed two cystic outpouchings of the meninges in the upper dorsal spine through minor defects in the dorsal vertebra filled with CSF with no evidence of cord compression and normal spinal cord and brain parenchyma without any evidence of hydrocephalus suggestive of two upper dorsal meningoceles. Radiography of the spine also showed lumbar vertebral defect suggestive of lumbar spina bifida occulta when correlated clinically.

In view of the above, patient was referred to pediatric neurosurgeon for further operative management. He was operated for it and had an uneventful post-operative recovery with no adverse neurological outcome and the patient is in follow-up.

DISCUSSION

Spinal dysraphism occurs in about 1-3/1000 live births. Extent and severity of neurological deficit depends on the degree of malformation of the neural placode and also the level of the defect (Venkataramana *et al.*, 2011). A lumbosacral defect is most common while cervical defects are the least frequent (Sarnat et al., 2006). There is usually an increasing neurological deficit as the defect extends higher into the thoracic region (Kinsman *et al.*, 2016). Neurological deficits include motor, sensory and sphincter dysfunction depending upon the severity and level. Cervical and thoracic meningoceles are usually not associated with hydrocephalus (Sarnat *et al.*, 2006). In occult dysraphism, spinal cord may be anchored to various tissues starting from skin, subcutaneous tissue, adipose tissue or cartilage causing neurological deficits.

The present case was unusual because the neural tube defect was present at three levels of the spinal cord; two upper dorsal meningoceles and a lumbar spina bifida occulta (seen clinically as a hirsute patch) and yet causing no neurological deficit. Apart for the asymptomatic multi-level presentation, another important feature was that the thoracic spine is itself a rare site of presentation when compared to lumbosacral, lumbar and thoracolumbar defects. The patient was however at risk of developing a CSF leak due to partially epithelialized skin over the meningocele prone for ulceration due to pressure effects.

Maternal folate deficiency, among others, is found to be an important causative factor in the occurrence of such defects. However, in this case, maternal folate levels done at the time of clinical presentation was found to be within normal limits.

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