A full-term, 2725-g female infant was born by vacuum extraction delivery following an uncomplicated pregnancy. The infant appeared normal except for a tuft of dark hair covering the skin of the lumbar area (Figure 1). She moved her extremities well and showed normal sensation and strength.

As part of the patient workup, a radiograph of the spine was obtained followed by imaging of the spine with ultrasonography in axial (Figure 2) and midsagittal planes (Figure 3) and magnetic resonance imaging of the spine (Figure 4).

From the Departments of Radiology (Drs Epelman and Golsher) and Neonatology (Drs Makhoul and Sujov), Rambam Medical Center, Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel. Dr Epelman is currently affiliated with the Department of Diagnostic Imaging, Hospital for Sick Children, Toronto, Ontario.
Denouement and Discussion

Occult Spinal Dysraphism (Tethered Cord, Diastematomyelia, and Spinal Lipoma)

The term dysraphism means incomplete fusion of a raphe. The skin and the nervous system share a common ectodermal origin. Differentiation of the neural ectoderm from the epithelial ectoderm occurs during the third to the fifth week of gestation. Cleavage occurs concurrently with formation and closure of the neural tube. The embryologic concurrence of events explains simultaneous occurrence of “occult” dysraphic lesions and minor malformations of the skin. Awareness of the cutaneous manifestations of underlying spinal cord malformations is critical because dysraphic malformations can cause damage from tethering of the spinal cord.

While during early fetal life the spinal cord occupies the entire length of the vertebral column, differential growth results in progressive ascent of the conus medullaris. In the newborn, the conus medullaris can be at the level of L2/L3 but by the age of 3 months is at the level of L1/L2, as in adults. Tethering of the cord prevents the progressive ascent of the conus with continued somatic growth and anchors the conus at or below the L2 level. Flexion of the normal spine is associated with upward movement of the cord and if the cord is fixed or tethered, traction injuries occur and there is damage to the lower cord and conus.

Split cord malformations are anomalies in which the spinal cord is cleft over a portion of its length by either a fibrocartilaginous or a bony septum originating from the posterior part of the vertebral body. The terms diastematomyelia and diplomyelia have created confusion; hence, they have been superseded by the term split cord malformation. The malformation results from disarray of midline axial integration during gastrulation, producing a split neural tube. In both types of malformation there is a tethering element that results in neurologic deterioration.

The neurologic symptoms result from flexion and extension movements of the cord that produce traction and trauma from the impaling septum. The clinical presentations of occult spinal lesions vary. Clinical signs could appear at birth or be delayed until adulthood. Tethering from any cause may produce back, leg, or groin pain, progressive lower extremity sensorimotor dysfunction, urinary dysfunction from neurogenic bladder, bowel incontinence or constipation, lower extremity orthopedic deformities, and progressive scoliosis.

Dysraphism should be diagnosed and surgically repaired before symptoms and signs of irreversible neurologic impairment are evident. Cutaneous abnormalities, such as hypertrichosis, a dimple, or a lipoma, might be the only clues to an underlying tethered cord. This is particularly seen in newborn infants with spinal dysraphism; 71% to 100% of patients have these signs. A high index of suspicion should occur when any of the above-mentioned cutaneous abnormalities is present so that spinal dysraphism can be recognized and managed to prevent irreversible neurologic impairment.

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Corresponding author: Monica Epelman, MD, Department of Diagnostic Imaging, Hospital for Sick Children, 555 University Ave, Toronto, Ontario, M5G 1X8, Canada.

REFERENCES


The Editor is seeking submissions for a new feature, Clinical Problem Solving, which will combine Picture of the Month, Radiological Case of the Month, and Pathological Case of the Month. Our aim is to demonstrate the thinking process of a master clinician involved in approaching a patient with an unknown disease. The discussion of such cases should place the clinician’s expertise into the context of the prevailing medical literature on the topic. Manuscripts should be between 3000 and 4000 words and may include photographs and radiographs.