Sonographic Findings in a Neonate With Diastematomyelia and a Tethered Spinal Cord

Po-Cheng Hung, MD, Huei-Shyong Wang, MD, Tai-Ngar Lui, MD, Alex M.-C. Wong, MD

Congenital midline cutaneous paraspinous lesions, mostly located in the lumbar sacral areas, are widely recognized as markers of occult spinal dysraphism. Diastematomyelia, or a split spinal cord, is an unusual congenital spinal abnormality in which the spinal cord is divided into 2 columns by a fibrous, cartilaginous, or bony septum. In most cases, lesions are located on the lower thoracic and upper lumbar parts of the spine. It often manifests in the neonatal period with midline paraspinous lesions. Sonography is a reliable tool for the study of the most severe spinal dysraphism. Diastematomyelia has been reported in neonates; however, to the best of our knowledge, the sonographic findings of diastematomyelia have rarely been described. We report a case of diastematomyelia and a tethered spinal cord in a neonate with lower back hemangioma at birth that was diagnosed via spinal sonography and was confirmed by spinal magnetic resonance imaging (MRI).

Case Report

A female neonate was the first child born to healthy, nonconsanguineous parents. There were no prenatal complications. The family history was noncontributory. The neonate was born by uncomplicated normal spontaneous vaginal delivery at 39 weeks' gestation. The Apgar scores were 9 at 1 minute and 10 at 5 minutes. Her birth weight was 2940 g (40th percentile); her length was 48.5 cm (50th percentile); and her occipitofrontal circumference was 34 cm (60th percentile). After birth, physical examination disclosed no abnormal findings except a hemangioma over the lower back at about the L2–L3 level (Figure 1). Neurologic examination showed no abnormal findings. Routine laboratory examinations revealed no abnormal findings. Screening results for toxoplasmosis, human immunodeficiency virus, measles, rubella, cytomegalovirus, herpes simplex, and other viruses were negative.
Diastematomyelia and a Tethered Spinal Cord

At the age of 2 days, spinal sonography was performed using an Acuson 128 XP ultrasound scanner (Siemens Medical Solutions, Mountain View, CA) equipped with a 7.0-MHz linear transducer. The sonogram (Figure 2) showed widening of the spinal canal with 2 hemicords in its own dural sheath and an additional echogenic focus between the 2 hemicords in the transverse plane. Coexisting tethering of the spinal cord was present in the longitudinal plane. Magnetic resonance imaging of the spine (Figure 3) showed complete separation of the spinal canal, thecal sac, and neural tissue at about the L2 level by a midline bony spur, a coexisting tethered spinal cord, and a dermal sinus tract. At 6 months old, a laminectomy was performed from L1 through L3, and the dura was opened around the hemangioma. The dorsal cartilage septum and ventral bony spicule between the 2 dural sacs were removed; the tethered spinal cord was detethered; and the dermal sinus tract was excised smoothly. The postoperative course was uneventful, and the patient's neurologic status remained normal at the 9-month follow-up.

Discussion

Cutaneous alterations as indirect signs of occult spinal dysraphism can be found in approximately 86% of instances. Cutaneous stigmata that may indicate underlying dysraphism of the lumbosacral spine include cutaneous dimples, pigment changes, nevi, hemangiomas, dermal sinuses, subcutaneous lipomas, other mesenchymal tumors, hair tufts, skin tags, and even tail-like cutaneous structures, the so-called human tail. Spinal dysraphism refers to a spectrum of congenital anomalies characterized by incomplete fusion of midline parenchymal, bony, and neural elements of the spine. Diastematomyelia is a rare but potentially devas-
tating spinal dysraphism classically characterized as a fibrous, cartilaginous, or bony septum separating 2 hemicords. In 1837, the term diastematomyelia was first used by Charles P. Ollivier to describe an abnormality of the spinal canal in which the dura is perforated by a bony spike or fibrous band to create 2 sleeves, each containing a portion of the spinal cord, that is divided into 2 parts sagittally, joining together above and below the spike. It is a part of split cord malformations, which are commonly divided into 2 types. Type 1 consists of 2 hemicords, each with its own dural sheath and separated by a rigid osseocartilaginous septum. This is the most common type. Type 2 has 2 hemicords housed in a common dural sheath separated by a nonrigid fibrous septum. The type 1 malformation is called diastematomyelia. The type 2 malformation is known as diplomyelia. The term diastematomyelia is used commonly to refer to both types of split cord malformations. Our patient belonged to the type 1 category. Diastematomyelia usually presents in childhood, but some patients are free of symptoms, and the condition may be discovered in adult life. The incidence or prevalence of diastematomyelia is not known. Girls have been previously reported in the majority. Diastematomyelia is most frequently found in the lumbar spine, followed by the thoracolumbar region and pure thoracic area. The spectrum of this anomaly progresses from simple splitting of the spinal cord to a cleft containing soft tissue to a cleft containing a fully developed bone. Diastematomyelia may be isolated, or it may be associated with abnormalities arising from various ectodermal or endodermal remnants such as spina bifida, kyphoscoliosis, butterfly vertebrae, hemivertebrae, a dermal sinus tract, and tethering of the spinal cord with lipoma, teratoma, dermoids, neurenteric cysts, and even meningocele, or myelomeningocele. Our case showed diastematomyelia with a dermal sinus tract and spinal cord tethering with lipoma.

The embryology of diastematomyelia remains unclear, although numerous theories have been proposed. Thompson et al proposed an aberrant event such as application of a teratogen on the primitive streak and node that form the notochord during the third week of embryogenesis, resulting in failure of separation of the neuroectodermal and endodermal layers. Cephadal growth of the notochord leads to a divergence at the level of the neuroectodermal adhesion, forming a split cord. Pang et al also suggested that the formation of adhesions between the ectoderm and endoderm lead to an accessory neurenteric canal between the yolk sac and amnion. Around this condenses an endomes-

Figure 3. A, Axial T2-weighted spinal MRI showing splitting of the spinal cord (arrow) by a bony spur (arrowhead). B, Sagittal MRI showing diastematomyelia (arrow) with spinal cord tethering by lipoma (arrowhead) and a dermal sinus tract (curved arrow).
enchymal tract that bisects the developing notochord, resulting in formation of 2 hemineural plates, leading to 2 cords. Because the cartilaginous ossification of the spinal processes in infants up to 12 months of age and the absence of posterior elements in dysraphism provide an "acoustic window" to the thecal space, spinal sonography has consistently been shown to be a useful technique in the assessment of congenital anomalies of the lower spine and is recommended as the first-line examination in neonates and young infants.12,13 Diastematomyelia has been diagnosed sonographically in utero.14 However, sonographic findings of diastematomyelia in neonates have rarely been described before.15–17 Transverse scans are essential for the sonographic diagnosis. In longitudinal views the abnormality may be overlooked because the 2 hemicords cannot be seen simultaneously. Glasier et al15 reported 2 cases with splitting of the cord within the spinal canal and spicule in 1 sonographically. Raghavendra et al16 reported a neonate with 2 hemicords within a single dural sheath visualized by spinal sonography. Bruhl et al17 also showed a spinal sonogram of a patient with a split cord with a bony spur in a single widened spinal canal. In our patient, the spinal sonogram showed widening of the spinal canal with 2 hemicords in its own dural sheath, an additional echogenic focus between the 2 hemicords in the transverse plane, and coexisting tethering of the spinal cord, which were confirmed via spinal MRI.

The prognosis is excellent if the septum is removed and the associated tethered spinal cord and dermal sinus tract are corrected in time.10 This is important because it has been found that neonates with no neurologic or orthopedic abnormality at birth may have progressive neurologic or orthopedic deficits later.

In conclusion, although diastematomyelia is a rare form of spinal dysraphism, cutaneous markers have a crucial role in the detection and diagnosis of occul spinal dysraphism and need further investigation when initially noted. Spinal sonography has consistently been shown to be a useful technique in the assessment of congenital anomalies of the lower spine and is recommended as the first-line examination in neonates and young infants.

References