A rare cutaneous sign of occult spinal dysraphism with tethered spinal cord

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A female, born after normal pregnancy and labor, was referred for consultation for a 1-cm long fibrous appendage in the midline of her lumbosacral region one day after birth (Fig. 1). Although she was neurologically intact, computed tomography at 3 months revealed L4-L5 spina bifida (Fig. 2A), and magnetic resonance imaging (MRI) showed a low conus medullaris level (L3), and a tethered dorsal spinal cord that seemed to connect with the site of the skin lesion (Fig. 2B). Spina bifida occulta with tethered spinal cord due to a dermal sinus tract was diagnosed and the patient was treated surgically. At operation, the sinus tract extending from the skin to the spinal cord was observed, but the thickening of the filum terminale was not evident.

Occult spinal dysraphism with tethered spinal cord, if not recognized, may result in permanent neurologic deficits. Moreover, the neurological disturbances can be difficult to demonstrate in a young baby and clinical evaluation of bladder function is equally difficult at this age. To prevent serious sequelae, early screening of high-risk infants with cutaneous markers that present in 50%-90% of patients, radiologic diagnosis using X-rays, either MRI or sonography, and urodynamic testing, and the appropriate timing of surgical untethering should be performed. However, dorsal cutaneous lesions may be ignored unless they are common lesions such as midline hemangiomas, hairy patches, dimples or sinuses, or pigmented macules. In order to screen effectively, the physician must have a high degree of suspicion for such a rare skin abnormality as this case.

References

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