A newborn with a skin lesion on the back

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A 24-year-old woman without a family history of congenital anomalies delivered her first baby after a minimally complicated prenatal course. Poor weight gain led to an ultrasound at 33 weeks gestation; this test and others at 13 weeks to determine gestational age and at 20 weeks to evaluate for anatomic abnormalities all had normal results.

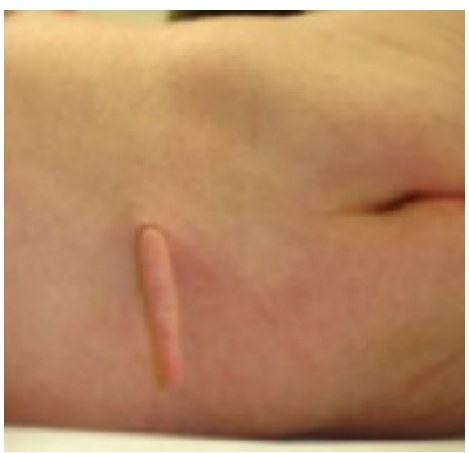
Prenatal screenings included a normal alpha feta-protein level. Review of the mother's medical records suggested compliance with routine prenatal care and vitamins; however, she smoked cigarettes during the pregnancy and occasionally took a pill containing butalbital, acetaminophen, and caffeine for migraine headaches. She did not have gestational diabetes, but late in the pregnancy she developed hypertension, necessitating induction of labor.

She delivered (via cesarean section) a full-term baby girl weighing 6 pounds, 9 ounces. The infant appeared healthy, with Apgar scores of 8 and 9 and a normal physical exam, except for a 2.5-cm appendage overlying the spine (**FIGURES 1 AND 2**). The 2-day nursery stay was that of a normal neonate, without complications. The pediatric surgeon who was consulted arranged for outpatient follow-up after hospital dismissal.

What is your diagnosis?

FIGURE 1

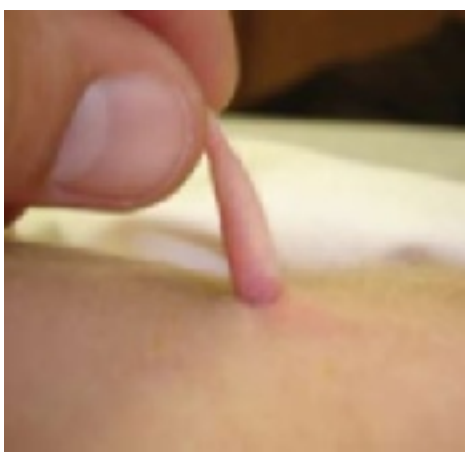
Skin lesion



The small appendage was in the middle of the infant's lower back.

FIGURE 2

Close-up



A close-up of the lesion, which contained primarily fatty tissue.

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FAST TRACK

When a tail-like structure is seen on a neonate, consider the possibility of occult spinal dysraphism before removing it

Diagnosis
Pseudotail

Inspection of the lesion at the time of excision revealed fatty tissue within its core, consistent with the diagnosis of pseudotail. Ultrasound of the spine performed before hospital discharge was normal.

Tails and pseudotails

Tails and pseudotails appear similar on physical exam and are differentiated histologically. True tails—remnants of the embryologic tail—contain a core of muscle fibers, nerves, and blood vessels, whereas pseudotails contain primarily fatty tissue.[1–3] From a practical and clinical point of view, you do not need to differentiate tails and pseudotails as they both can be associated with skin-covered congenital anomalies of the spine, or occult spinal dysraphism (OSD).

Although tail-like structures rarely occur, when they are present you should consider OSD.[3] Two noteworthy literature reviews support this. The first identified 33 tails and pseudotails, of which 10 had OSD.[1] An analysis of more recent cases (1960 through 1997) demonstrated OSD in 29 of 59 cases with lesions described as tails.[2] A higher incidence of OSD occurred between 1980 and 1997 in this series. The authors attribute the higher incidence to the advent of computed tomography and magnetic resonance imaging (MRI), thereby stressing the importance of spine imaging with the presence of any caudal appendage resembling a tail.

Epidemiology of occult spinal dysraphism

The true incidence of OSD is not known. Congenital anomalies of the central nervous system, second only to birth defects of the cardiovascular system, are reported on birth certificates, but OSD may go unrecognized in the neonatal period. Frequently a benign-appearing midline cutaneous lesion is the only evidence of OSD at birth.[4] Although 3% to 8% of newborns with a skin lesion over the spine have OSD,[5] 50% to 90% of patients with OSD have a skin marker.[5–7] Of 2010 newborns in 1 study, 144 had a midline cutaneous lesion, and 5.5% of these had an abnormal ultrasound suggesting OSD.[8] Additionally, the presence of more than 1 lesion increases the probability of finding OSD.[5,9,10]

You should look for underlying OSD in newborns with a suggestive skin lesion, since the embryologic origin of the spinal cord and the skin overlying the spine are the same: the ectoderm layer of cells within the trilaminar (endoderm, mesoderm, ectoderm) early embryo gives rise to both structures. Midline cutaneous lesions with high likelihood for OSD include hypertrichosis (hairy patch), atypical dimple (a dimple >5 mm diameter or >2.5 cm above the anus), hemangioma, lipoma, aplasia cutis, dermoid cyst or sinus, skin tags, tails, and pseudotails.[3]

Evaluation
The neonate with a midline cutaneous lesion

The incidence of OSD appears low in newborns with midline cutaneous lesions, but irreversible orthopedic, urologic, and neurologic problems from OSD may occur later in life. These complications can be prevented by early surgical intervention; therefore, it is critical that you do not miss a diagnosis of OSD and pursue spine imaging.

The definitive test, an MRI,[3,6,9] usually requires sedation of infants,[11] making high-resolution ultrasound the recommended choice for initial screening.[3–10] Ultrasound is not invasive and may allow for testing in the hospital nursery. After age 3 months, when ossification of the posterior spine limits visualization of the spinal cord by ultrasound, consider an MRI first. If the ultrasound demonstrates OSD or is inconclusive, follow with an MRI for a more specific diagnosis.

Indications for an ultrasound of the

newborn's spine include any one of the following: an abnormal antenatal ultrasound; a midline cutaneous lesion other than a simple dimple; presence of other OSD related anomalies; or an abnormal neurologic exam.[5] The authors of this protocol retrospectively applied it to 223 infants who had an ultrasound, and it reduced the number of tests by 50% without missing a single case of OSD.

If you are caring for a newborn infant you should resist the temptation to simply remove a midline cutaneous lesion, including a pseudotail, in the nursery without first searching for OSD. If OSD is present, neurosurgical consultation should be obtained as soon as possible. The optimal time for surgical intervention is within the first 4 months of life.[4]

Patient outcome

The infant's pseudotail was removed without consequence and postoperatively the patient has thrived.

CONFLICT OF INTEREST

The authors do not have any conflict of interest related to this manuscript.

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Photographs contributed by Jason G. Nirgiotis, MD.

FAST TRACK

High-resolution ultrasound is recommended for the initial screening of OSD