Cutaneous manifestations of split cord malformations

Report of three cases

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A split cord malformation (SCM) is a rare congenital disorder involving a division of the spinal cord that results in two distinct hemicords divided by bone, cartilage, or a fibrous septum. Cutaneous manifestations of SCMs are common, including hypertrichosis, dimples, hemangiomas, nevi, lipomas, and sinus tracts. In this report, the authors describe cutaneous manifestations of SCMs in three patients, including a rare report of near fully formed digits overlying the SCM. (DOI: 10.3171/PED-07/09/240)

KEY WORDS • atretic meningocele • cutaneous manifestation • pediatric neurosurgery • spinal lipoma • split cord malformation

Case Reports

Case 1

This 3-month-old boy presented with a congenital area of abnormal skin in the midline at the level of L-2, consistent with the characteristics of an atretic meningocele (Fig. 1). The patient’s development had been normal, as were the results of a neurological examination. No preoperative diagnostic studies were performed. At operation, the meningocele was excised and an underlying fibrous tract was followed through the interspinous ligament. The tract terminated in a bone spur that perforated the dural sac in the midline. The fibrous tract and spur were excised and the child made an uneventful recovery.

Case 2

This 5-year-old girl presented with a congenital midthoracic deformity. Her neurological examination results were normal. The deformity consisted of two immobile fingers that exited from a caudal, subcutaneous lipoma (Fig. 2A). A radiograph demonstrated bones in the digital appendages and the midline bone spur (Fig. 2B). At operation, the digits were surgically removed. The metacarpal-like bones at the base of the fingers were contiguous with the bone spur dividing the split cord. The lipoma and bone spurs were excised (Fig. 2C). The postoperative course remained uneventful and the child recovered without neurological deficits.

Case 3

This 7-year-old girl with no neurological deficits presented with an upper thoracic skin-covered lesion (Fig. 3A). Bone was palpable in the pointed, cephalic aspect of the lesion. A computed tomography scan demonstrated a bone spur dividing the spinal cord and an overlying cyst of mixed contents (Fig. 4). At operation, the palpable subcutaneous cutaneous manifestations of split cord malformations, formerly termed diastematomyelia, refer to a subset of double spinal cord malformations. Diastematomyelia was first described in 1837 by Ollivier, and the Greek derivation references the word diastema, meaning cleft, and myelos, referring to the spinal cord. The cleft is usually found in the lower thoracic or upper lumbar regions. The resultant hemicords usually contain distinct central canals with intact pia mater, and often form a separate dorsal and ventral horn. Type I SCMs result in an osseocartilaginous median septum dividing the hemicords, resulting in two separate arachnoid and dural sleeves. Type II SCMs traverse a single subarachnoid space within a single dural sleeve divided by a fibrous septum. The hemicords usually reunite caudal to the split. We report three cases involving typical and atypical cutaneous signs of SCMs encountered during the senior author’s (A. L. A.) trip to Kenya; this author also performed all surgeries involved in these cases.

Abbreviation used in this paper: SCM = split cord malformation.
bone spur was excised first (Fig. 3B). The cyst was dissected from adjacent tissue and found to contain mucinous material consistent with a neurenteric cyst. The cyst was excised and the intraspinal bone spur was resected (Fig. 3C). The patient had no postoperative neurological deficits. A postoperative cerebrospinal fluid leak developed and was repaired with no further sequelae.

**Discussion**

Split cord malformations represent a dysraphic state of unknown embryogenesis. The prevailing theory regarding the pathogenesis of SCMs involves the initiation of this condition by an accessory neurenteric canal. This canal initially connects the yolk sac to the amnion via the primitive knot. The knot normally migrates caudally to the coccyx. During migration, an accessory canal may develop, splitting the neural ectoderm to form a midline fistula between the neural canal and notochord, enabling continued contact between the ectoderm and endoderm within the canal. Once the neural plate is split, mesenchymal tissue may fill the gap resulting in an endomesenchymal tract. This tissue has been postulated to originate from the vertebral bodies. Recent surgical models involving chick embryos have lent credence to these theories, demonstrating that a resorbable agar screen does not induce an SCM, whereas an implanted membrane shell or notochord does induce the formation of a split cord. These results provide evidence in opposition to the aberrant primary neurulation hypothesis of SCM. Instead, noninvolution of a firm midline structure prevents fusion and allows infiltration of mesodermal cells from the notochord. The timing of this endomesenchymal tract and the resultant recruitment of precursor cells from the meninx primitiva may determine the formation of either a single dural sleeve (Type II SCM) or intervening septum (Type I SCM).

The theory readily explains the diverse set of cutaneous manifestations found in SCMs, as these manifestations are derived from residual mesodermal elements of the endomesenchymal tract. Cutaneous manifestations occur in more than half of patients with SCM and commonly include focal hirsutism, nevi, telangiectasias, atrophic skin, lipomas, dermal sinuses, and capillary hemangiomas. As reviewed in this paper and consistent with the literature, most skin overlying SCMs remains intact, implying early closure of the accessory neurenteric canal, which allows the ectoderm to form intact skin. Hypertrichosis, followed by capillary hemangioma, is considered to be the best predictor of an underlying SCM, occurring in as many as 56% of patients in one series, although these conditions are commonly found together. The focal hypertrichosis may include a “faun’s tail,” manifesting as a coarse, hairy, round or oval patch several inches in length. Other hair anomalies include “silky down,” represented by smaller areas of fine, nonterminal, or lanugo hair, or simply focal hypertrichosis consisting of hair with a normal appearance developing along the midline.

Common vertebral anomalies include spina bifida, kyphoscoliosis, butterfly vertebra, and hemivertebra. The bone abnormality may become evident as a subcutaneous and hard bone prominence, as seen in Case 3. Others have reported this phenomena, detailing two patients with palpable, midline, bone protuberances that were covered by intact skin. Surgical extirpations revealed these protrusions to be isolated subcutaneous bone nodules and lipomas with overlying bone laminae. Other associated SCM anomalies include tethered cords, teratomas, inclusion dermoids, lipomas, syringohydromy-
elia, and Chiari malformations. Even more rare anomalies, as seen in Case 2, include tail-like lipomas. Tavafoghi and colleagues reported a single case of a 21-year-old female patient who presented with a 4 × 1.3–cm tapering, tail-like structure emanating from the midline at L-5. This appendage was soft and covered by normal skin with numerous fine hairs. Imaging revealed a low tethered cord with an SCM. The intraoperative details of this report were short and only briefly noted the tail-like structure’s connection to a subcutaneous lipoma. Yamada and associates detailed a similar finding, more similar to our own findings, of a dorsal midline proboscis found on a 2-year-old girl in the upper lumbar area. The appendage was 7.5 × 1 cm, triply segmented, and manifested three fingerlike growths. Surgical exploration revealed a hemilipomyelomeningocele and tethered cord associated with a Type II SCM. The appendage in Case 2 contained fingernails and an almost anatomic reproduction of the bony architecture of the phalanges. The only prior citation of an SCM associated with near anatomical phalanges was in 1889, in which Jones and Larkin reported the removal of an accessory limb contiguous with a meningocele and bifurcated spinal cord. This supernumerary limb contained three fingers and arose from the midline cervicothoracic junction. Previous reports detailing more anatomical manifestations of accessory limbs or digits with associated spinal cord dysraphism have not been associated with SCMs. These reports detail accessory appendages associated with lipomyelomeningoceles or sacrococcygeal teratomas.

Conclusions

In this paper, we report three cases of cutaneous manifestations of SCMs. These findings contribute to the accumulated knowledge regarding cutaneous manifestations of split cord abnormalities that should alert physicians and caregivers to the underlying pathology involved.
References


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