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Case Report

Cervical wart-like cutaneous appendage with a contiguous stalk of limited dorsal myeloschisis treated with untethering after long-term follow-up

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ABSTRACT

Background: Limited dorsal myeloschisis (LDM) is a condition in which the separation of the neuroectoderm from the cutaneous ectoderm during primary neural tube formation results in localized disjuncture, causing a continuous cord-like connection and spinal cord tethering. We reported a case of cervical LDM with a wart-like cutaneous appendage that was treated with excision after long-term follow-up.

Case Description: The patient was an 18-year-old girl. A wart-like cutaneous appendage was noted over the nape of the neck since birth. Computed tomography showed spina bifida in the cervical and thoracic spines, and spinal magnetic resonance imaging (MRI) showed a cervical skin lesion and an enlarged dural sac in the dorsal thoracic spinal cord. At 18 years of age, the patient occasionally experienced numbness in her left hand and was referred to our outpatient clinic due to a new high signal intensity in the dorsal cervical spinal cord on a T2-weighted MRI. The MRI showed that a cord-like object was continuous intradural and dorsal to the spinal cord from a cutaneous lesion in the median cervical region, with a high signal in the same region. Symptomatic cervical spinal cord tethering due to a cord-like material was diagnosed, and the patient underwent resection. During surgery, the tract was removed from the cutaneous lesion into the dura mater as a single mass and untethered in the dorsal spinal cord. The histological diagnosis was a pseudo-dermal sinus tract with no luminal structures or neural tissue present, as the cord-like substance was connective tissue containing small blood vessels. Based on the neuroimaging and pathological findings, the patient was diagnosed with cervical LDM. Neurological symptoms improved postoperatively.

Conclusion: Herein, we reported a case of cervical LDM that was treated after long-term follow-up. The patient's symptoms improved immediately after surgery. Cervical LDMs are rare, and the timing of surgery for LDM should be considered according to the patient's condition.

Keywords: Limited dorsal myeloschisis, Spinal dysraphism, Tethered cervical cord

INTRODUCTION

Limited dorsal myeloschisis (LDM) was first described as a distinct clinicopathological condition by Pang *et al.*[11] LDMs are characterized by two invariable features: a focal closed neural tube defect and a fibroneural stalk linking the skin lesion to the spinal cord.[11,12] Embryogenesis of

LDM is hypothesized to be an incomplete disjunction between the cutaneous and neural ectoderms.[11,12] In all LDMs, the fibroneural stalk starts at the skin lesion and is tethered to the spinal cord; the recommended treatment involves untethering the stalk from the spinal cord.[11,12]

LDMs are categorized based on their skin manifestations as saccular and non-saccular.[11,12] Saccular LDM consists of a skin-based cerebrospinal fluid sac topped by a squamous epithelial dome, whereas non-saccular LDM has a flat squamous epithelial surface or a sunken crater or pit.[7,8,11,12] We treated a patient with cervical LDM and a wart-like cutaneous appendage.

Given its rarity, reports on long-term outcomes in patients with LDM are limited. Here, we describe a case of cervical LDM that became symptomatic; the patient was treated after 18 years of long-term follow-up.

CASE PRESENTATION

An 18-year-old girl was referred to our hospital for left-hand numbness. She was born at 33 weeks and 6 days of gestation and weighed 1800 g at birth. A wart-like cutaneous appendage with dimples was observed over the nape of the neck. Since there were no abnormal neurological findings, the patient was placed under observation. Neuroradiological examinations were performed at 1 year of age. Spina bifida was found in the cervical (C1–C5) and thoracic (T3–T5) spine on spinal computed tomography [Figure 1a]. Furthermore, the magnetic resonance imaging (MRI) showed a small dimple in the cervical skin lesion and an enlarged subarachnoid space in the dorsal thoracic spinal cord [Figure 1b]. As there were no neurological symptoms, the patient was kept under observation. During the observation period, she underwent regular follow-ups with MRI at a nearby hospital [Figures 1c-e]. At 18 years of age, the patient occasionally experienced numbness in her left hand and was referred to our hospital due to a new high-signal intensity in the dorsal cervical spinal cord on T2-weighted MRI [Figure 2a]. MRI showed a cord-like object continuous intradural and dorsal to the spinal cord from a cutaneous lesion in the median cervical region, with a high signal in the same region [Figure 2b]. A wart-like cutaneous appendage was observed on the nape of the neck [Figure 2c].

Symptomatic cervical spinal cord tethering due to the cord-like material was diagnosed, and the patient underwent resection with monitoring of the somatosensory-evoked potential. A skin incision was made around the warts. We carefully dissected the tract and followed it all the way down to its penetration of the dura [Figure 3a]. The dura was opened at the midline above the tract. The intradural portion ended by attaching to the dorsal surface of the spinal cord with thickened arachnoid tissue [Figure 3b]. The entire tract

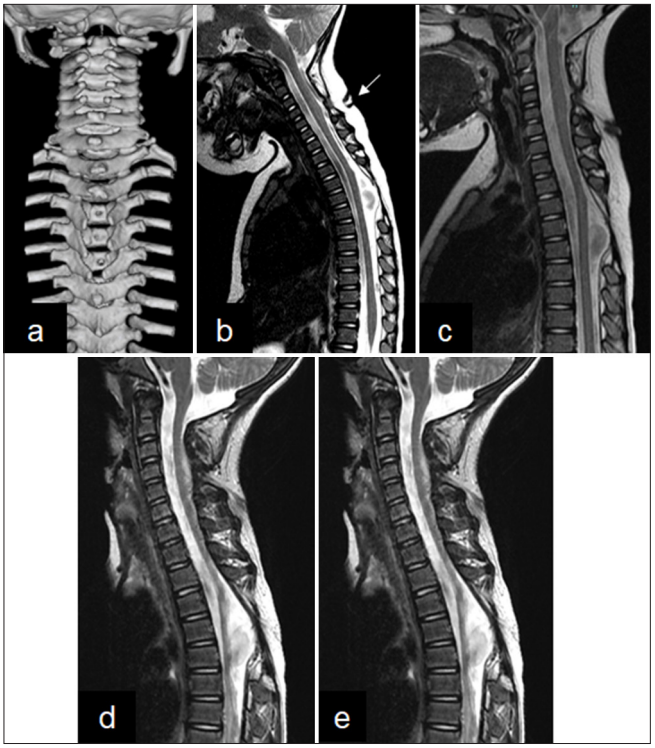

Figure 1: Neuro-radiological imaging during follow-up. (a) Computed tomography showed spina bifida at the cervical (C1–C5) and thoracic (T3–T5) spine. (b) Mid-sagittal T2-weighted MRI of cervical spine at age 2. The arrow indicated the skin lesion. (c-e) Mid-sagittal T2-weighted MRI of the cervical spine at age 2 (c), age 10 (d), and age 14 (e). No obvious high–signal-intensity in the cervical spinal cord. MRI: Magnetic resonance imaging.

with the wart-like cutaneous appendage was excised after the thickened arachnoid tissue tethered to the spinal cord was severed to release the spinal cord [Figure 3c] fully.

Postoperatively, the patient's symptoms improved and did not present any new neurological abnormalities. MRI showed that the tethered spinal cord was released, and the high-signal lesion was reduced 6 months after surgery [Figure 3d]. The enlarged dural sac located at the thoracic spina bifida remained asymptomatic and, therefore, did not require treatment.

Histopathologically, the epidermal ridge was composed of a mixture of subcutaneous fat and connective tissue [Figures 4a and b]. A cord-like structure composed of connective tissue containing small blood vessels extending from the subcutaneous fat tissue was observed [Figures 4c and d]. No luminal structures, cutaneous structures, or neural tissues were observed. Elastica van Gieson staining revealed that the cord-like structures were composed of collagen fibers [Figure 4e], and there was no evidence of nerve tissue or nerve sheaths stained for glial fibrillary acidic protein or S-100 protein. The histological diagnosis was a pseudo-dermal sinus tract.

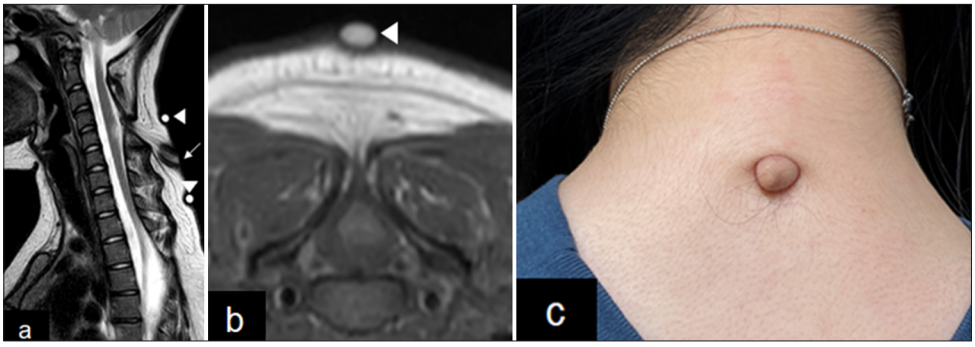

Figure 2: Neuroradiological and photo-imaging of skin lesion. (a) Mid-sagittal T2-weighted MRI of cervical spine at age 18. The arrow indicated skin lesions, and arrowheads were landmarks of the skin lesion at the time of MRI examination. (b) Axial T2-weighted MRI at C5. The spinal cord is tethered posteriorly to the tract. The arrowheads are landmarks of the skin lesion at the time of MRI examination. (c) Photograph showing a wart-like cutaneous appendage over the nape of the patient's neck. MRI: Magnetic resonance imaging.

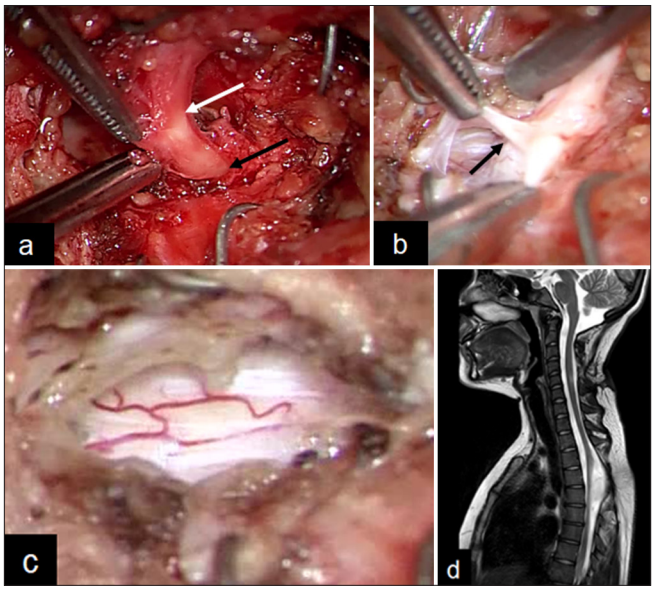

Figure 3: Operative photographic views and MRI 6 months after surgery. (a) The subcutaneous stalk was continuous into the dura mater (white arrow: Subcutaneous tract; black arrow: Dural penetration). (b) The intradural tract is connected to the dorsal spinal cord (black arrow). (c) The intradural tract is untethered from the spinal cord. (d) Mid sagittal T2-weighted MRI of cervical spine 6 months after surgery. The high-signal legion was reduced at the dorsal cervical spine.

Based on the neuroimaging and pathological findings, the patient was diagnosed with cervical LDM.

DISCUSSION

In this case, cervical LDM caused a tethered cord during adulthood. However, congenitally tethered cervical spinal cords are extremely rare in adults. In a few reported cases, it has been associated with a dermal sinus that enters the subarachnoid spaces and blends with dorsal spinal cord elements.[2,5,9,12] Ackerman and Menezes reviewed 30 years of experience with spinal congenital dermal sinuses in 28 patients.[1] The locations were as follows: the cervical region in five patients, a thoracic region in four patients, a lumbar region in nine patients, and below or at the lumbosacral junction in ten patients. Among all the patients, only four were adults. Two of these patients had cervical LDMs. Moreover, three of the four adult patients experienced pain due to a tethered cord.

In the case of the dermal sinus, the tract leads from the epidermis for a variable distance through the dermis, subcutaneous fat, fascia, muscle, vertebral arch, and meninges up to the spinal cord. Of these layers, only the epidermis and spinal cord are ectodermal in origin. Histological analysis of a typical dermal sinus tract revealed that this fistula had a lumen lined by stratified squamous epithelium immediately surrounded by dermal tissue.[15]

In this case, the cord-like structure was comprised of connective tissue containing small blood vessels and no luminal structures. Based on these pathological findings, the patient was diagnosed with LDM.

Cervical non-saccular LDMs are not commonly observed. Pang *et al*. described the distribution of cases by the types of LDM.[12] In their case series, over two-thirds of the LDMs were located in the lower half of the spinal cord. The flat or crater LDMS were found in the lumbar and lower thoracic regions, and saccular types were found in both cervical and lumbar segments. Maroufi *et al*. reported 22 cases of saccular cervical LDMs.[3] In this series, the age of the patients at the time of admission ranged from 13 days to 10 years (mean ± standard deviation: 19.40 ± 36.03 months). Of the 22 patients, eight had neurological deficits. Moreover, Sarukawa *et al*. reported two patients with LDM with a human tail-like cutaneous appendage in lumbosacral or sacral lesions.[14] Our patient

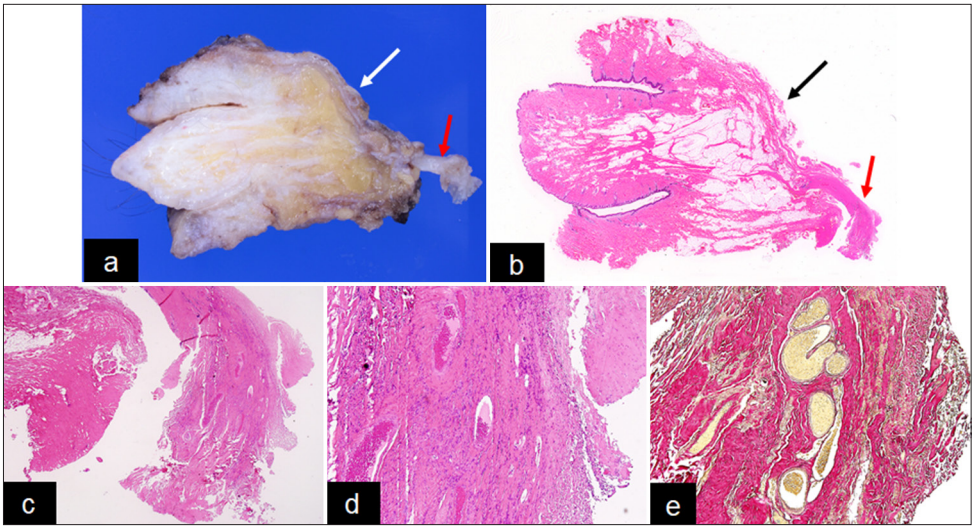

Figure 4: Histopathological findings of the resected specimens. (a) Macroscopic image of the split surface of appendage (white arrow: Appendage, red arrow: Tract). (b) Macroscopic image of hematoxylin and eosin (H&E) stained appendage (black arrow: Appendage, red arrow: Tract). The appendage contained mature fibroadipose tissue. The tract, composed of connective tissue containing small blood vessels, extends from within the subcutaneous fat tissue (H&E: ×12.5 (c), ×40 (d)). (e) Elastica van Gieson staining revealed that the tracts were composed of collagen fibers (×40).

had a cutaneous appendage on her neck, which was a rigid mass that did not contain cerebrospinal fluid. Furthermore, the appendage was dome-shaped rather than tail-shaped. Wart-like LDM is uncommon and has not yet been reported.

Retrotraction and fixation of the cervical spinal cord seem to play important roles in flexion and extension movements in a relatively fixed spinal cord. This may explain the onset of symptoms in the adult age, where stretching and tension on neural tissues can occur with impairment of spinal circulation, leading to metabolic dysfunction, neuronal injury, and progressive neurological impairment.[2,13,17]

The patient underwent regular outpatient visits and MRI examinations. There were no symptoms during follow-up, and there was no abnormal signal in the spinal cord. However, when she experienced numbness in her left hand, a new high-signal-intensity lesion was found in the dorsal cervical spinal cord on T2-weighted MRI.

MRI is a noninvasive method used to monitor the pathological features of spinal cord lesions. Several histopathological studies of cervical spinal cord injury have reported that a blurred intramedullary high-signal intensity on T2-weighted images is thought to represent edema or petechial hemorrhage.[10]

Several studies have reported the preoperative relevance of MRI in patients with cervical spondylotic myelopathy. Morio *et al*. reported that patients with altered signal intensity on both T1- and T2-weighted images demonstrated the worst postoperative prognosis compared with those who showed a preoperative signal change only on T2-weighted images.[6] Furthermore, Mastronardi *et al*. reported that patients with cervical spondylotic myelopathy with signal intensity changes only on T2-weighted images showed better prognosis if there was regression of signal hyperintensity on the follow-up MRI.[4] Although our patient had a cervically tethered spinal cord, her symptoms promptly improved after surgery, and a regression of signal hyperintensity was observed on the follow-up MRI. Even in the absence of symptoms, it is important to perform periodic MRIs to determine the time of operation. When there is only hyperintensity on T2-weighted images, surgery should be performed before the hypointensity appears on T1-weighted images.

In this case, the patient was concerned about the cosmetic aspects of the neck lesion and numbness. During surgery for LDM with an appendage, untethering of the cord and cosmetic removal of the appendage should be performed during the same operation, as is typical for other tethered lesions with an appendage.[16]

CONCLUSION

Cervical LDMs are rare, and wart-like cutaneous appendages rather than saccular lesions are even rarer. Clinicians should be aware of possible morphological variations in skin lesions associated with LDM.

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