Child's Nervous System (2019) 35:973–978
https://doi.org/10.1007/s00381-019-04071-w

ORIGINAL ARTICLE

Human tail-like cutaneous appendage with a contiguous stalk of limited dorsal myeloschisis

Mio Sarukawa[1] · Takato Morioka[2] · Nobuya Murakami[2] · Takafumi Shimogawa[3] · Nobutaka Mukae[3] · Noriko Kuga[4] · Satoshi O. Suzuki[5] · Koji Iihara[3]

Received: 8 January 2019 / Accepted: 20 January 2019 / Published online: 6 February 2019

Abstract

Purpose Limited dorsal myeloschisis (LDM) is characterized by a fibroneural stalk linking the skin lesion to the underlying spinal cord. On account of the external skin lesion, all LDMs are either flat (nonsaccular) or saccular, and a human tail-like cutaneous appendage has not been reported.

Methods In our 14 LDM patients, 2 had tail-like appendages. We retrospectively analyzed the relationship between the appendage and the LDM tract from the clinicopathological findings of these 2 patients.

Results Preoperative magnetic resonance imaging including three-dimensional heavily T2-weighted images demonstrated an intradural tethering tract, but failed to reveal the precise communication with the appendage. However, surgery revealed the extradural and intradural slender stalk, starting at the base of appendage and running through the myofascial defect. Histological examination demonstrated that there was a tight anatomical relationship between the fibroadipose tissue of the appendage and the fibrocollagenous LDM stalk.

Conclusion When there is potential for an LDM stalk in patients with an appendage, a meticulous exploration of the stalk leading from an appendage is required. Clinicians should be aware of possible morphological variations of skin lesions associated with LDM.

Keywords Glial fibrillary acidic protein · Fibroneural stalk · Untethering · Fibroadipose tissue

Introduction

Limited dorsal myeloschisis (LDM) was first described as a distinct clinicopathological entity [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]. The embryogenesis of LDM is hypothesized to be 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 cord, and recommended treatment consists of untethering the stalk from the cord [11, 12].

LDMs are categorized based on their skin manifestations as saccular and nonsaccular (flat) [11, 12]. Saccular LDM consists of a skin-based cerebrospinal fluid (CSF) sac topped by a squamous epithelial dome, while flat LDM has a squamous epithelial flat surface or a sunken crater or pit [8, 9, 11, 12]. Recently, we treated 2 LDM patients with a human tail-like cutaneous appendage as an external skin manifestation. We analyzed the relationship between the appendage and the LDM tract from the clinicopathological findings of these 2 patients.

Methods

From July 2015 to December 2018, 14 Japanese LDM patients underwent initial untethering surgery at Kyushu University Hospital and a related hospital by the supervision

✉ Takato Morioka
takato@ns.med.kyushu-u.ac.jp

[1] Department of Pediatric Neurology, Fukuoka Children's Hospital, Fukuoka, Japan

[2] Department of Neurosurgery, Fukuoka Children's Hospital, 5-1-1 Kashii-teriha, Higashi-ku, Fukuoka 813-0017, Japan

[3] Department of Neurosurgery, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

[4] Department of General Pediatrics & Interdisciplinary Medicine, Fukuoka Children's Hospital, Fukuoka, Japan

[5] Department of Neuropathology, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

of the senior author (TM). As an external skin manifestation, 8 patients had flat lesions and 4 saccular lesions. Two patients had tail-like appendages. We retrospectively analyzed the clinical, neuroradiological, intraoperative, and histopathological findings of these 2 patients to demonstrate the relationship between the appendage and the LDM tract. Preoperative magnetic resonance imaging (MRI), including three-dimensional (3D) T1-weighted spoiled gradient-recalled echo image and 3D heavily T2-weighted image (3D-hT2WI), as well as conventional T1- and T2-weighted images, was performed with a 1.5 T MRI machine, as previously described [7–10].

Results

Both patients (case 1 and case 2) were boys and born by normal delivery. Their neurological examinations were normal. However, both were noted to have appendages in the lumbosacral or sacral lesions situated just off the midline to the right side (Figs. 1a and 2a). In case 2, hypertrichosis was also noted around the appendage (Fig. 2a).

At 3 months of age, MRI, in both cases, especially 3D-hT2WI (slice thickness, 1.25 mm), demonstrated an intradural tethering tract originating at the dural wall of the lower lumbar level and joining the lower-lying conus at the L3 level (Figs. 1b, c and 2b, c). However, in case 1, MRI failed to reveal the communication between the extradural stalk and the appendage (Fig. 1d). In case 2, a slender band of isointensity, starting at the subcutaneous region and entering the intraspinal canal, was faintly observed on 3D-hT2WI (Fig. 2b), while the precise anatomical relationship with the appendage was not evident (Fig. 2d).

At 4 months of age, in both cases, surgery consisting of untethering of the cord and cosmetic removal of the appendage was performed during the same operation. A lipomatous stalk started at the base of appendage and passed through the myofascial defect below the L5 level in case 1 (Fig. 1e-1, f-1) and the bifid S2 level in case 2 (Fig. 2e-1). Laminoplastic laminotomy revealed a slender stalk (1.0 mm diameter in case 1; 1.3–1.4 mm diameter in case 2) that was contiguous to the subcutaneous lipomatous stalk, and which ran epidurally and entered intradurally at the L4 level in case 1 (Fig. 1e-2, f-2) and S1 level in case 2 (Fig. 2e-2, e-3, f-2). After opening the dura, the intradural stalk was found to be continuous with the extradural stalk (Figs. 1e-3, f-3 and 2e-3, f-2, f-3). In case 1, the intradural stalk was found to be joined to the dorsal surface of the lower-lying conus at L2–3 (Fig. 1e-3, f-3). In the vicinity of the stalk–cord attachment, the diameter of the stalk was 2.0 mm. The intradural stalk was then disconnected from the cord. In case 2, the stalk was severed at the most rostral part of the operative field, and the cord was untethered (Fig. 2e-4). The proximal cut end of the stalk receded deep into the spinal canal. In both cases, the intradural and extradural stalks and appendage with the contiguous lipomatous stalk were removed and submitted for histological examination (Figs. 1e-4 and 2e-4).

Postoperatively, our cases did not develop any neurological deficits. Histopathologically, in both cases, the appendage consisted of mature fibroadipose tissues covered by skin tissue (Fig. 3a, h). The subcutaneous lipomatous stalk consisted of fibroadipose tissues, which were contiguous to the fibroadipose tissue of the appendage, and were continuous with the fibrocollagenous tissues of the epidural stalk (Fig. 3b-d, i, j). The extradural (Fig. 3e, k) and intradural (Fig. 3f, k) stalks included fibrocollagenous tissues embedded with peripheral nerve fibers (Fig. 3g) and dilated vessels (Fig. 3h). No glial fibrillary acidic protein (GFAP)-immunopositive neuroglial tissues were observed in the stalks of the both cases.

Discussion

The central histopathological feature of an LDM stalk is GFAP-immunopositive neuroglial tissue in the fibrocollagenous band, a hallmark of the stalk's origin from the non-disjoined neuroectoderm [4–6, 8, 9, 11, 12]. However, no GFAP-immunopositive neuroglial tissue was noted in our case. Our previous reports [8, 9] demonstrated that detection of sparse GFAP-immunopositive neuroglial tissue is difficult with routine histopathological examination, since small islands of these tissues might well be missed during sectioning of the stalk. The diagnosis of LDM should be made based on comprehensive evaluation of other histological findings including the presence of peripheral nerve fibers, as observed in our patient, as well as clinical manifestations [8, 9]. Peripheral nerve fibers in that context have to come from neural crest cells that were present in the lateral dorsal tips of the coapting neural plates during the final stages of primary neural tube closure. Focal incomplete disjunction between the cutaneous and neural ectoderms might well pick up these neighboring neural crest cells and thus include their outgrowths of peripheral nerves into the stalk [8, 9]. In our case, furthermore, there was a tight anatomical relationship between the appendage and the stalk through the myofascial defect. The intervention of fibroadipose tissue does not contradict the LDM diagnosis, as our previous histological reports [4, 8, 9] demonstrated fibroadipose tissue between the skin lesion and stalk in many LDM patients, indicating that there is not always a tight fibrous connection.

However, preoperative MRI failed to reveal this connection, which may relate to the small size of the stalk. In the present case, we confirmed the diagnostic ability of 3D-hT2WI for detection of the intradural stalk, as 3D-hT2WI provides excellent contrast between the stalk and CSF [8–10, 14]. However, it is difficult to visualize a slender stalk

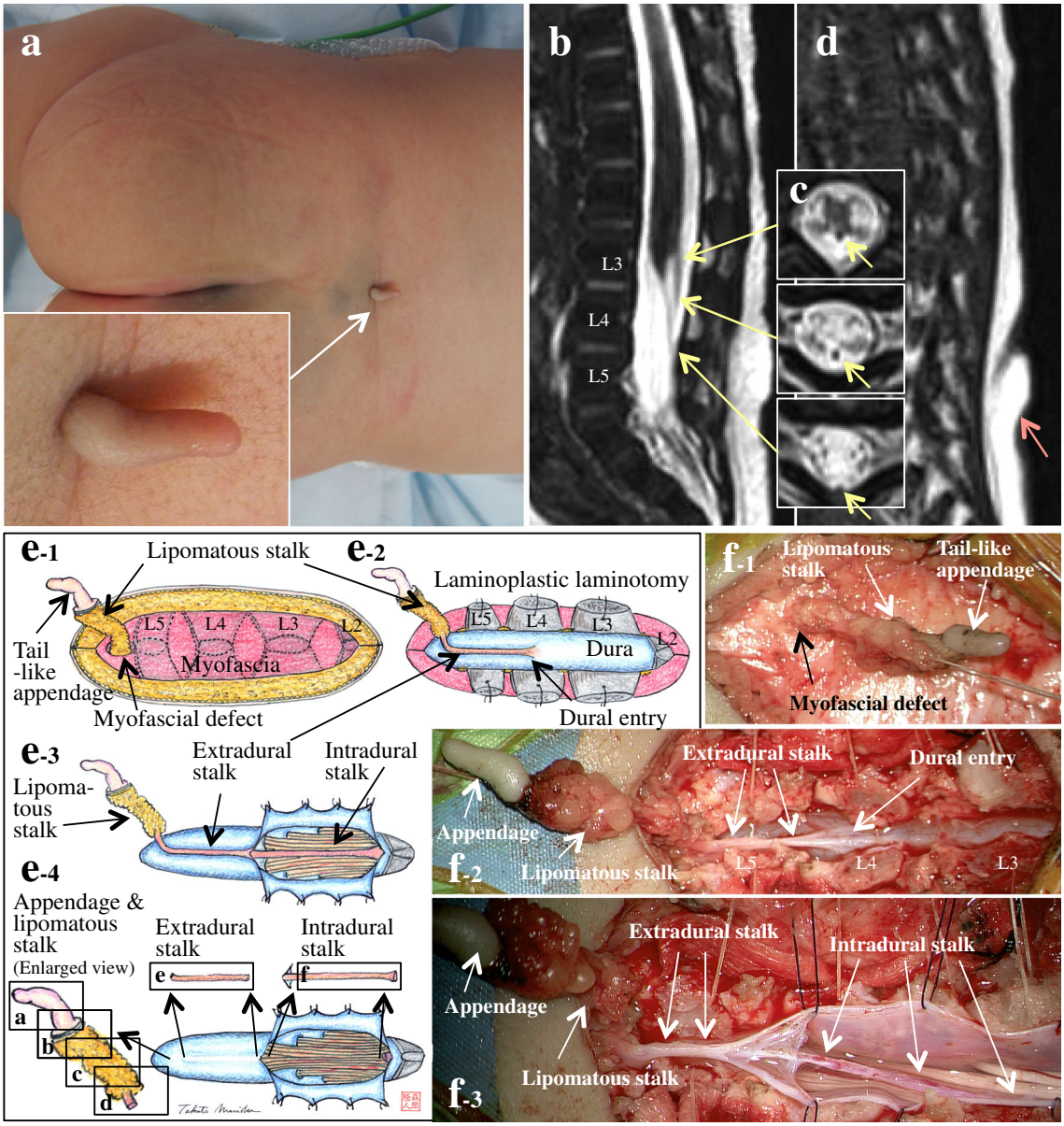

Fig. 1 Case 1. (**a**) Photograph showing a human tail-like cutaneous appendage (white arrow). The appendage is situated in the lumbosacral region just off the midline to the right side. Inset indicates the enlarged view of the tail. Sagittal view of three-dimensional heavily T2-weighted images (slice thickness, 1.25 mm) (**b**) and serial axial views of T2-weighted images (slice thickness, 3.90 mm) (**c**) show an intradural tethering stalk (yellow arrows) starting at the L4–5 level and joining the lower-lying conus at the L3 level. (**d**) Sagittal view of three-dimensional T1-weighted spoiled gradient-recalled echo image (slice thickness, 1.25 mm) fails to reveal an extradural stalk continuous with the appendage (indicated with red arrow). (**e**) Schematic drawing and (**f**) microscopic view of the operative findings. (**e-1, f-1**) An elliptical skin incision is made around the appendage, and the subcutaneous lipomatous stalk at the base of the appendage is dissected out. The lipomatous stalk passes through the myofascial defect below the L5 level. (**e-2, f-2**) Following laminoplastic laminotomy at L3–5, a slender stalk with a diameter of 1.0 mm contiguous to the lipomatous stalk was found to run epidurally and enter intradurally at the L4 level. (**e-3, f-3**) Upon opening the dura, the intradural stalk was found to join the dorsal surface of the lower-lying conus at L2–3. In the vicinity of the stalk–cord attachment, the diameter of the stalk is 2.0 mm. (**e-4**) The intradural stalk is disconnected from the dura and the cord to untether the cord. The intradural and extradural stalks and appendage with the contiguous lipomatous tract are then removed and submitted for histological examination

outside of the CSF space [8, 9]. The 1.25-mm-thick sections obtained by 3D-hT2WI in the present cases still provided insufficient details of the slender stalk with a diameter of only 1.0–1.4 mm. It is also possible that the subcutaneous lipomatous stalk was composed of fibroadipose tissue, which is indistinguishable from subcutaneous fibroadipose tissue on MRI.

Dao and Netsky originally proposed a two-category classification for human tails: (1) true tails and (2) pseudotails. In this classification, true tails are vestigial remnants of

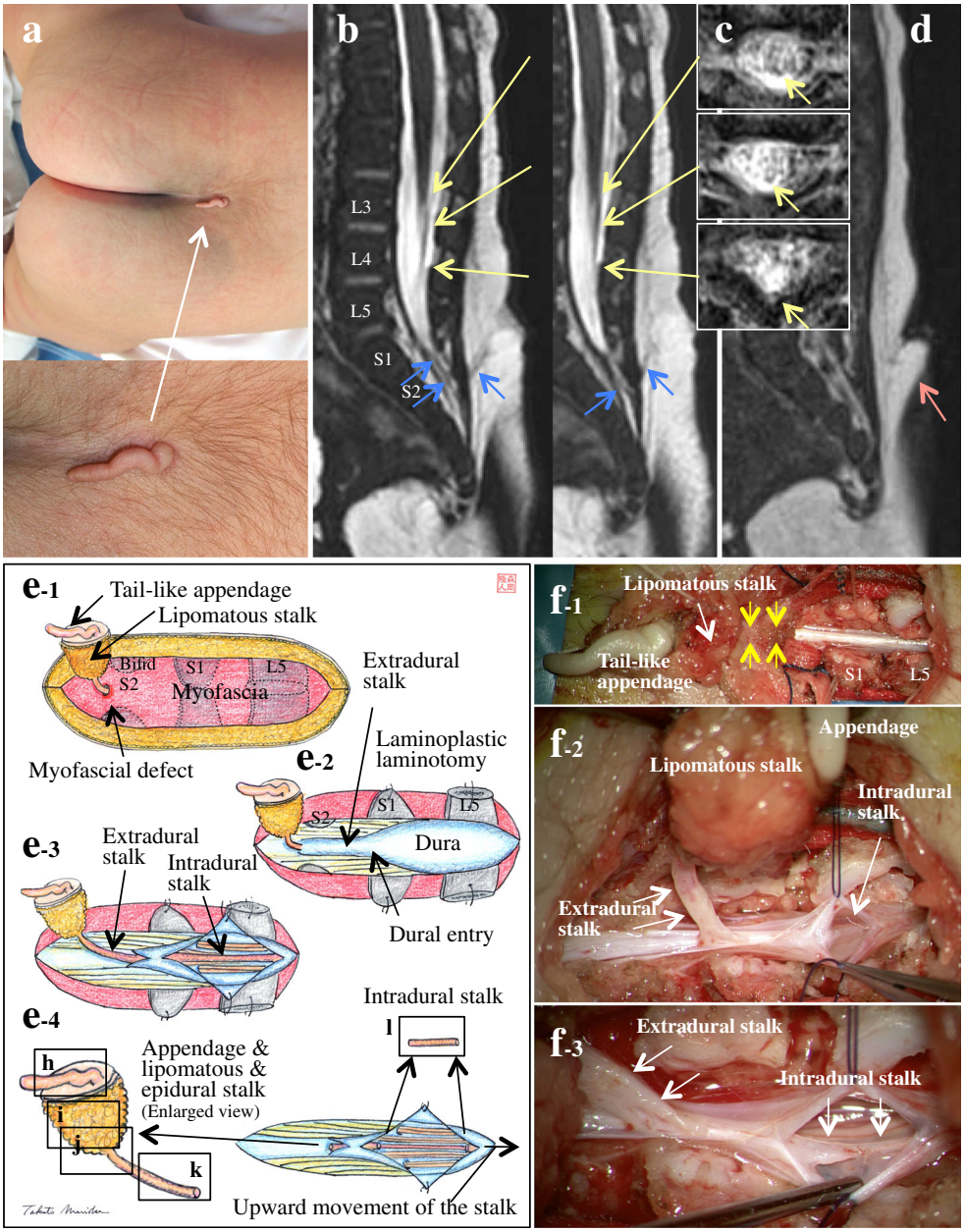

Fig. 2 Case 2. (**a**) Photograph showing an appendage (white arrow). The appendage is situated in the sacral region just off the midline to the right side. Inset indicates the enlarged view of the appendage. Hypertrichosis is noted around the appendage. Serial sagittal views of three-dimensional heavily T2-weighted images (slice thickness, 1.25 mm) (**b**) and serial axial views of T2-weighted images (slice thickness, 3.90 mm) (**c**) show an intradural tethering stalk (yellow arrows) starting at the L5 level and joining the lower-lying conus at the L3 level. A slender band of isointensity (indicated with blue arrows) starting at the subcutaneous region and entering the intraspinal canal are faintly observed. (**d**) Sagittal view of three-dimensional T1-weighted spoiled gradient-recalled echo image (slice thickness, 1.25 mm) fails to reveal an extradural stalk continuous with the appendage (indicated with red arrow). (**e**) Schematic drawing and (**f**) microscopic view of the operative findings. (**e-1**) The subcutaneous lipomatous stalk at the base of the appendage is dissected out. The lipomatous stalk passes through the myofascial defect at the bifid S2 level. (**e-2, f-1, f-2**) Following laminoplastic laminotomy at L3–5, a slender stalk (indicated with yellow arrows on f-1) with a diameter of 1.3–1.4 mm contiguous to the lipomatous stalk was found to run epidurally and enter intradurally at the S1 level. (**e-3**) Upon opening the dura, the intradural stalk was found to be continuous with the extradural stalk. (**e-4**) The intradural stalk is disconnected from the dura and the cord to untether the cord. The extradural stalk and appendage with the contiguous lipomatous tract are then removed and submitted for histological examination

embryonic human tails, whereas pseudotails are skin manifestation of underlying abnormal structures such as lipomas or teratomas [1]. However, recent studies have made this system outdated [15–17], and it is now generally accepted that tails are not remnants of the embryonic tails, but rather cutaneous markers for various spinal tethering lesions, including spinal

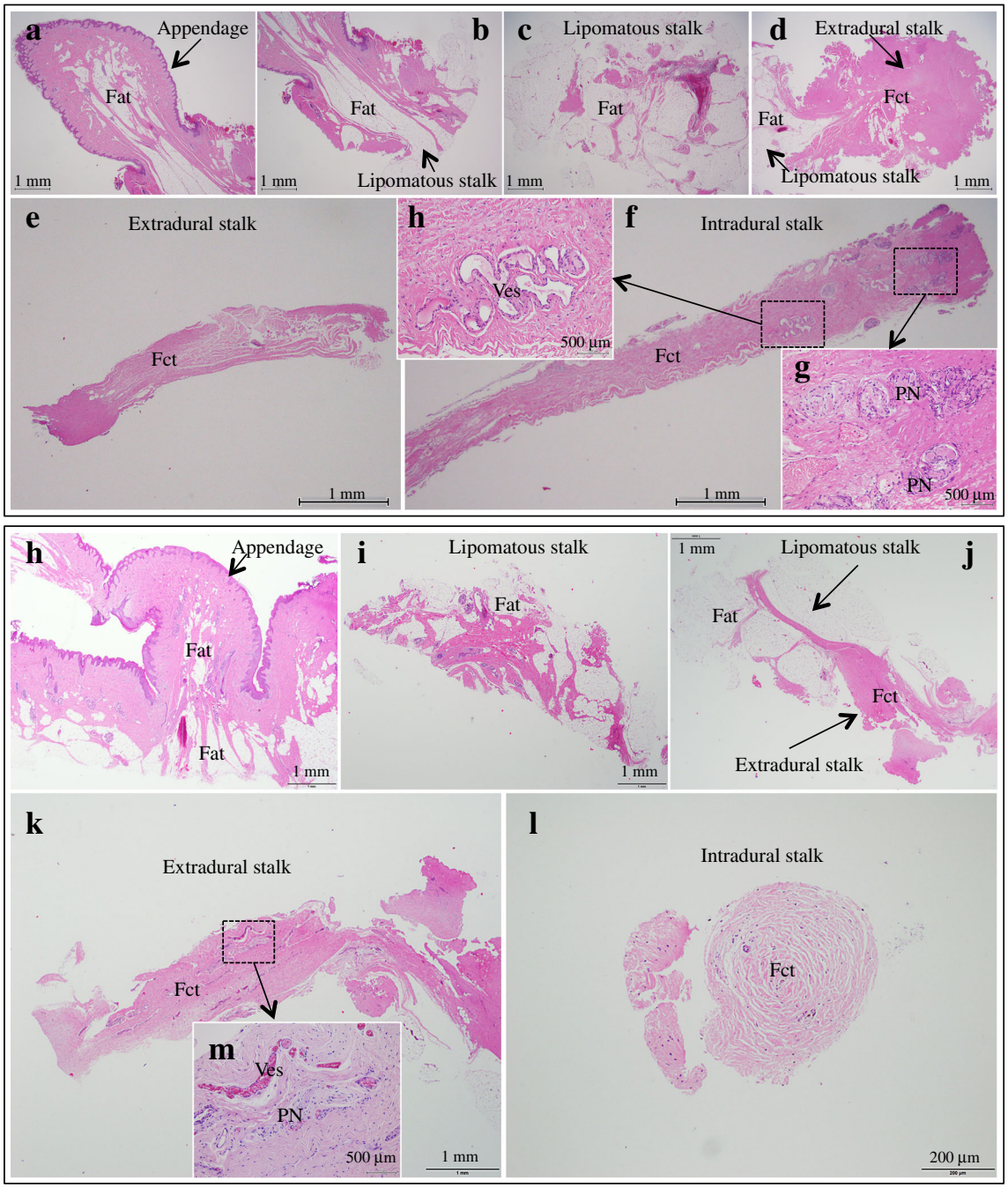

Fig. 3 Histopathological findings of the resected specimens in case 1 and case 2. Each section is indicated as an open square in Fig. 1e-4 and Fig. 2e-4, respectively. (**a-g**) Case 1. (**a**) The appendage of case 1 consists of mature fibroadipose tissue (Fat) covered by skin tissue, which had a finely jagged squamous epithelium. (**b-d**) The subcutaneous lipomatous stalk is composed of fibroadipose tissue, which is contiguous to the appendage, and continuous with the fibrocollagenous tissue (Fct) of the epidural stalk. Longitudinal sections of the extradural (**e**) and intradural (**f**) tract showing fibrocollagenous tissue embedded with peripheral nerve fibers (PN, g) and large vessels (Ve, h). (g, h) High-power views of the area are indicated by the doted open square in (**f**). (h-l) Case 2. (**h**) The appendage of case 2 also consists of mature fibroadipose tissue covered by skin tissue. (**i, j**) The subcutaneous lipomatous stalk is composed of fibroadipose tissue, which is contiguous to the appendage, and continuous with the fibrocollagenous tissue of the epidural stalk. Longitudinal section of the extradural (**e**) and cross section of the intradural (**f**) tract showing fibrocollagenous tissue embedded with peripheral nerve fibers and large vessels

lipomatous malformation, dermal sinus tracts, and split cord malformation [2, 7, 13, 15–17]. When such a tethering lesion is present, the appendage is frequently connected to it through dysraphic posterior elements [17]. However, an LDM stalk in connection with the appendage has not been reported. Wilkinson and Boylan [17] reported that 2 of 8 appendages

in their series had tracts with central neurovascular or neural cores, and discussed the relationship with the neuroectodermal appendages proposed by Gaskill and Merlin [3]. In this malformation, they speculated that a fibroneural stalk extends outward and forms a tail-like structure [3, 17].

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 tethering lesions with an appendage [17]. An issue with preoperative diagnosis is that the slender LDM stalk is difficult to recognize even with 3D-hT2WI [8, 9, 14]. Consequently, if there is potentially an LDM stalk, a meticulous exploration of the stalk leading from an appendage is required [17]. In conclusion, clinicians should be aware of possible morphological variations of skin lesion associated with LDM.

Acknowledgments We thank Drs. Nobuko Kawamura, Ayumi Tsukamoto, and Ryutaro Kira for supporting our study. We also thank Edanz Group (www.edanzediting.com/ac) for editing a draft of this manuscript.

Funding information This work was partly supported by the Research Foundation of Fukuoka Children's Hospital.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Dao AH, Netsky MG (1984) Human tails and pseudotails. Hum Pathol 15:449–453
2. Donovan DJ, Pedersen RC (2005) Human tail with noncontiguous intraspinal lipma and spinal cord tethering: case report and embryologic discussion. Pediatr Neurosurg 41:35–40
3. Gaskill SJ, Marlin AE (1989) Neuroectodermal appendages: the human tail explained. Pediatr Neurosci 15:95–99
4. Hiraoka A, Morioka T, Murakami N, Suzuki SO, Mizoguchi M (2018) Limited dorsal myeloschisis with no extradural stalk linking to a flat skin lesion: a case report. Childs Nerv Syst 34:2497–2501
5. Lee JY, Chong S, Choi YH, Phi JH, Cheon J-E, Kim S-K, Park SH, Kim I-O, Wang K-C (2017) Modification of surgical procedure for "probable" limited dorsal myeloschisis. J Neurosurg Pediatr 19:616–619
6. Lee JY, Park S-H, Chong S, Phi JH, Kim S-K, Cho B-K, Wang K-C (2019) Congenital dermal sinus and limited dorsal myeloschisis: "Spectrum disorders" of imcomplete dysjunction between cutaneous and neural ectoderms. Neurosurgery 84:428–434
7. Morioka T, Murakami N, Shimogawa T, Mukae N, Hashiguchi K, Suzuki SO, Iihara K (2017) Neurosurgical management and pathology of the lumbosacral lipomas with tethered cord. Neuropathology 37:385–392
8. Morioka T, Suzuki SO, Murakami N, Shimogawa T, Mukae N, Inoha S, Sasaguri T, Iihara K (2018) Neurosurgical pathology of limited dorsal myeloschisis. Childs Nerv Syst 34:293–303
9. Morioka T, Suzuki SO, Murakami N, Mukae N, Shimogawa T, Haruyama H, Kira R, Iihara K (2019) Surgical histopathology of limited dorsal myeloschisis with flat skin lesion. Childs Nerv Syst 35:119–128
10. Murakami N, Morioka T, Hashiguchi K, Yoshiura T, Hiwatashi K, Suzuki SO, Nakamizo A, Amano T, Hata N, Sasaki T (2013) Usefulness of three-dimensional T1-weighted spoiled gradient-recalled echo and three-dimensional heavily T2-weighted images in preoperative evaluation of spinal dysraphism. Childs Nerv Syst 29:1905–1914
11. Pang D, Zovickian J, Oviedo A, Moes GS (2010) Limited dorsal myeloschisis: a distinctive clinicopathological entity. Neurosurgery 67:1555–1580
12. Pang D, Zovickian J, Wong S-T, Hou YJ, Moes GS (2013) Limited dorsal myeloschisis: a not-so-rare form of primary neurulation defect. Childs Nerv Syst 29:1459–1484
13. Samura K, Morioka T, Hashiguchi K, Yoshida F, Miyagi Y, Yoshiura T, Suzuki SO, Sasaki T (2009) Coexistence of a human tail and congenital dermal sinus associated with lumbosacral lipoma. Childs Nerv Syst 25:137–141
14. Tomita Y, Morioka T, Murakami N, Noguchi Y, Sato Y, Suzuki OS (2018) Slender stalk with combined features of saccular limited dorsal myeloschisis and congenital dermal sinus in a neonate. Pediatr Neurosurg. https://doi.org/10.1159/000495810
15. Tubbs RS, Malefant J, Loukas M, Oakes WJ, Oskouian RJ, Fries FN (2016) Enigmatic human tails: a review of their history, embryology, classification, and clinical manifestations. Clin Anat 29:430–438
16. Turk CC, Kara NN, Bacanli A (2016) The human tail: a simple appendage or cutaneous stigma of an anomaly? Turk Neurosurg 26:140–145
17. Wilkinson CC, Boylan AJ (2017) Proposed caudal appendage classification system; spinal cord tethering associated with sacrococcygeal eversion. Childs Nerv Syst 33:69–89