Cervicothoracic Limited Dorsal Myeloschisis: An Under-Recognized Potential Mimicker

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Consent
Yes, written informed consent was obtained from the patient for the publication of this case report and accompanying images

Ethical Statement / Human and animal rights
Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Conflict of Interest
All the authors have indicated they have no financial relationships relevant to this article to disclose.

ABSTRACT

Limited dorsal myeloschisis results from incomplete disjunction during spinal cord development, leading to a persistent stalk between skin and spinal cord. Neurological symptoms in limited dorsal myeloschisis patients vary depending on the level of spinal cord involvement. Diagnosis typically relies on cutaneous markers and magnetic resonance imaging findings, which can differentiate limited dorsal myeloschisis from other conditions like dermal sinus or myelomeningocele. Sometimes medullary edema/local pre syrinx potentially caused by this entity can exhibit a pseudo tumoral aspect and mimic other differential diagnoses such as tumors and inflammatory/demyelinating conditions. Surgical intervention involves resection of the stalk to alleviate tethering effects, with newer techniques aimed at minimizing invasive procedures while ensuring complete removal of the stalk.

CASE REPORT

BACKGROUND

This case highlights a rare presentation of oculomotor nerve palsy due to neurovascular compression by a fetal posterior cerebral artery (PCA), a finding that remains underreported in the radiology literature. While neurovascular conflicts are well-documented for the trigeminal and facial nerves, isolated oculomotor nerve involvement is uncommon, with limited documented cases. Advanced high-resolution MR imaging (3D CISS, FIESTA, and T2-SPACE) and MRA played a crucial role in identifying the neurovascular conflict, emphasizing their value in cases of unexplained cranial neuropathies. This report also underscores the clinical relevance of pupillary sparing, which may lead to misdiagnosis if neurovascular compression is not considered. Furthermore, the case provides insights into recurrent, progressive oculomotor neuromyotonia (ONM), likely influenced by pulsatile compression and ischemic effects. Recognizing this entity is essential for radiologists and clinicians to avoid misattributing oculomotor palsy to more common causes such as ischemia or demyelination. By reinforcing the importance of detailed neurovascular evaluation, this case contributes to both diagnostic accuracy and patient management strategies.

CASE REPORT

A 37-year-old female patient has been followed up for an unclear diagnosis of transverse myelitis, likely a post-viral syndrome or demyelinating disorder for twelve years. During the initial presentation, she presented with left upper extremity numbness/tingling. She was oriented and with no motor deficits. She has not had any further neurological attacks. The work up with multiple blood tests including inflammatory markers and paraneoplastic panel was negative, except for a positive finding of high Kappa Lamba ratio of 1.99 (normal range 0.26 to 1.65), which has normalized overtime. The remainder of her blood tests, including immunoglobulins and electrophoresis, were normal. There was also no evidence of hematological disorder. In addition, no skin stigmata were identified on the patient's back.

Imaging findings

Neuroimaging with C-spine magnetic resonance imaging (MRI) (Figures 1,2) showed long-term stability of a focal enlargement of the C7-T1 spinal cord with abnormal signal predominantly in the left lateral and posterior columns, resembling a pseudo-tumoral appearance. At the same level, there was a tiny focal disruption of the posterior dura matter, contiguous to the spinal cord through a fibrous rudimentary tissue (neural stalk). Features compatible with congenital non-saccular limited dorsal myeloschisis with local pre syrinx/spinal edema. After multidisciplinary discussion, the neurology team thinks that her previous neurological symptoms were not due to this lesion itself. Therefore, neuroimmunology follow-up was no longer suggested.

DISCUSSION

Etiology & Causes

Limited dorsal myeloschisis (LDM) is a closed spinal dysraphism due to incomplete disjunction between cutaneous and neural ectoderms during primary neurulation [1], with a persistent stalk which prevents complete midline skin closure. This entity can be classified as saccular or non-saccular (flat).

The stalk can exhibit intradural, extradural and subcutaneous portions, and it may not all be seen depending on its composition, length and thickness may not be identified [2,3]. Regarding the composition, peripheral nerve fib00000000ers and smooth muscle fibers of neural crest origin may be dragged into the stalk during incomplete disjunction, while the striated muscle fibers of mesodermal origin may enter the stalk along with the lipomatous tissues during premature disjunction. This can also have glial fibrillary acidic protein (GFAP)-immunopositive neuroglial tissues. The tract, depending on its cellular constituents and its completeness in extent, can lead to tethered cord syndrome and/or collection of dermal sinus tissue with their dreadful effects (Figure 3). Rarely, if there is an associated inflammatory process, nodular or longitudinal enhancement can be seen. There are only few case reports describing it on the cervical segment [3].

Clinical findings

Many patients with LDM present at a young age. Approximately half of these patients exhibit no neurological deficits at the time of diagnosis in infancy, highlighting the significance of cutaneous markers as an initial diagnostic indicator. The characteristic cutaneous marker for both flat and saccular LDMs is a localized area of abnormal epithelium along the dorsal midline [2].

It's important to note that other cutaneous markers of dysraphism, such as hypertrichosis, capillary hemangioma, or a misaligned gluteal crease, are never seen alone in LDM cases without the essential epithelial crater or pit. LDMs cause neurological deficits exclusively through their tethering effect, with the type and severity of these deficits depending on the spinal level affected by the LDM. Overall, patients with LDM typically experience milder disabilities compared to those with other forms of dysraphic malformations, such as split cord malformations and spinal cord lipomas [2].

The proximity of tension to the relevant spinal cord segments appears to correlate with the type of deficits observed. For instance, only cervical lesions cause hand and arm weakness; leg weakness is found in only 9% of cervical lesions, but increases to 22% in upper thoracic lesions, 38% in thoracolumbar lesions, and 50% in lumbar lesions. Bladder dysfunction occurs in approximately 15% of lower thoracic and lumbar lesions, but is not seen in cervical or upper thoracic lesions. Lumbar LDMs near the conus are particularly concerning as they more frequently affect the bladder and are often more difficult to detect. Similar to other conditions involving spinal cord tethering, the likelihood of neurological injury increases with the longitudinal growth of the spine and with age [2].

Imaging Findings

MRI is the imaging technique of choice to delineate the details of pathological anatomy of the malformation especially regarding composite lesions [2]. The neuroimaging findings of LDM follow embyrogenetic logic and illustrate precisely the various anatomic forms. The defining feature present in all LDMs is the fibroneural stalk connecting the base of the cutaneous lesions to the underlying spinal cord following a discrete extraspinal and then intrathecal course. The entire ‘‘tract’’ is usually traceable on MRI [4]. However, in this case, there was no appreciable component of the tract along the posterior soft tissues to the skin, suggesting a very thin and/or obliterated tract.

Although it is known that the cord to which the stalk attaches is enlarged [5] and mimics an intramedullary pseudo-tumoral lesion, depending on the stalk thickness and orientation, this entity can be easily missed on neuroimages, as the presented case. A high index of suspicion from the neuroradiologist is important for assessing these long-term stable findings and optimizing the protocol with thin slices to confirm them.

Differential Diagnoses

The stalk is usually obliterated, and the skin may have variable coverage, and a cutaneous stigma is not mandatory. These characteristics help differentiate it from a congenital dermal sinus, in which it is not obliterated and there is no intact skin covering the defect, with the presence of a cutaneous stigma/dimple. These 2 conditions can even coexist [2].

From the neuroimaging standpoint, the medullary edema/local pre-syrinx potentially caused by this entity can exhibit a pseudo-tumoral aspect and mimic other differential diagnoses such as tumors, especially astrocytoma, and inflammatory/demyelinating conditions including transverse myelitis and multiple sclerosis.

Treatment & Prognosis

The surgical indication of an LDM is well established and involves relatively straightforward resection of the stalk to untether the cord. A modification of the 0surgical technique has also been suggested to avoid long segment laminectomies, but still enables complete excision of long LDM stalks after identification of its dural entry and its merge point with the cord. In this particular case, surgical resection of the LDM stalk was not performed because there was no definitive neurological correlation with the MRI findings, and clinical follow-up was preferred.

TEACHING POINT

Limited dorsal myeloschisis (LDM) presents with cutaneous markers along the dorsal midline and can lead to neurological deficits due to spinal cord tethering. Understanding the clinical and imaging features of LDM is crucial for accurate diagnosis and appropriate management, which typically involves surgical resection of the tethering stalk to alleviate symptoms and prevent further neurological complications.

QUESTIONS

Applies to article: *Murakami N, Morioka T, Suzuki SO et al. Clinicopathological findings of limited dorsal myeloschisis associated with spinal lipoma of dorsal-type. Interdisciplinary Neurosurgery 21 (2020) 100781. https://doi.org/10.1016/j.inat.2020.100781.*

1 - Which of the following statements is true about limited dorsal myeloschisis causes?

a) There is no persistent stalk in limited dorsal myeloschisis.

b) Can be classified as saccular or non-saccular.

c) It has a complete fusion between cutaneous and neural ectoderms during primary neurulation.

d) It has a complete disjunction between cutaneous and neural ectoderms during primary neurulation.

e) Is an opened spinal dysraphism.

Explanation:

a) There is no persistent stalk in limited dorsal myeloschisis [a persistent stalk which prevents complete midline skin closure.]

b) Can be classified as saccular or non-saccular (applies).

c) It has a complete fusion between cutaneous and neural ectoderms during primary neurulation. [It has an incomplete disjunction between cutaneous and neural ectoderms during primary neurulation.]

d) It has a complete disjunction between cutaneous and neural ectoderms during primary neurulation. [It has an incomplete fusion between cutaneous and neural ectoderms during primary neurulation.]

e) It is an opened spinal dysraphism. [Limited dorsal myeloschisis is a closed spinal dysraphism.]

Applies to article: *Wong ST, Pang D. Focal Spinal Nondisjunction in Primary Neurulation: Limited Dorsal Myeloschisis and Congenital Spinal Dermal Sinus Tract. J Korean Neurosurg Soc. 2021 Mar;64(2):151-188. doi: 10.3340/jkns.2020.0117.*

2 - Which of the following statements is true about limited dorsal myeloschisis?

a) The stalk of limited dorsal myeloschisis can exhibit just intradural and subcutaneous portions.

b) One third of the patients exhibit no neurological deficits at the time of diagnosis.

c) Cutaneous markers can be an initial diagnostic indicator.

d) Cutaneous marker is not a localized area of abnormal epithelium along the dorsal midline.

e) Hypertrichosis, capillary hemangioma, or a misaligned gluteal crease are not cutaneous marker of limited dorsal mieloschisis. Is an opened spinal dysraphism.

Explanation:

a) The stalk of limited dorsal myeloschisis can exhibit just intradural and subcutaneous portions. [The stalk can exhibit intradural, extradural and subcutaneous portions.]

b) One third of the patients exhibit no neurological deficits at the time of diagnosis. [half of these patients exhibit no neurological deficits at the time of diagnosis]

c) Cutaneous markers can be an initial diagnostic indicator. (applies)

d) Cutaneous marker is not a localized area of abnormal epithelium along the dorsal midline. [Cutaneous marker for both flat and saccular limited dorsal myeloschisisis a localized area of abnormal epithelium along the dorsal midline.]

e) Hypertrichosis, capillary hemangioma, or a misaligned gluteal crease are not cutaneous marker of limited dorsal mieloschisis. [Hypertrichosis, capillary hemangioma, or a misaligned gluteal crease, are never seen alone in limited dorsal myeloschisis cases without the essential epithelial crater or pit]

Applies to article: *Wong ST, Pang D. Focal Spinal Nondisjunction in Primary Neurulation: Limited Dorsal Myeloschisis and Congenital Spinal Dermal Sinus Tract. J Korean Neurosurg Soc. 2021 Mar;64(2):151-188. doi: 10.3340/jkns.2020.0117.*

3 - Which of the following statements is true about limited dorsal myeloschisis?

a) Patients with limited dorsal mieloschisis typically experience harder disabilities compared to those with other forms of dysraphic malformations

b) Bladder dysfunction occurs in approximately 50% of lower thoracic and lumbar lesions

c) Only dorsal lesions cause hand and arm weakness

d) Limited dorsal mieloschisis causes neurological deficits exclusively through their tethering effect.

e) Leg weakness is found in only 90% of cervical lesions

Explanation:

a) Patients with limited dorsal mieloschisis typically experience harder disabilities compared to those with other forms of dysraphic malformations. [patients with limited dorsal

myeloschisis typically experience milder disabilities compared to those with other forms of dysraphic malformations.]

b) Bladder dysfunction occurs in approximately 50% of lower thoracic and lumbar lesions. [Bladder dysfunction occurs in approximately 15% of lower thoracic and lumbar lesions.]

c) Only dorsal lesions cause hand and arm weakness. [Only cervical lesions cause hand and arm weakness]

d) Limited dorsal mieloschisis causes neurological deficits exclusively through their tethering effect. (applies)

e) Leg weakness is found in only 90% of cervical lesions. [Leg weakness is found in only 9% of cervical lesions]

Applies to articles: *Wong ST, Pang D. Focal Spinal Nondisjunction in Primary Neurulation: Limited Dorsal Myeloschisis and Congenital Spinal Dermal Sinus Tract. J Korean Neurosurg Soc. 2021 Mar;64(2):151-188. doi: 10.3340/jkns.2020.0117 and Pang D, Zovickian J, Oviedo A, Moes GS. Limited dorsal myeloschisis: a distinctive clinicopathological entity. Neurosurgery. 2010 Dec;67(6):1555-79; discussion 1579-80. doi: 10.1227/NEU.0b013e3181f93e5a.*

4 - Which of the following statements is true about limited dorsal myeloschisis?

a) CT is the imaging technique of choice to delineate the details of pathological anatomy of the malformation especially regarding composite lesions.

b) The neuroimaging findings of limited dorsal myeloschisis follow evolutionary logic

c) The neuroimaging findings do not illustrate the various anatomic forms.

d) The defining feature presents in all LDMs is the fibroneural stalk connecting the base of the cutaneous lesions to the underlying spinal cord following a discrete extraspinal and then intrathecal course

e) The entire ‘‘tract’’ is usually traceable on CT.

Explanation:

a) CT is the imaging technique of choice to delineate the details of pathological anatomy of the malformation especially regarding composite lesions. [MRI is the imaging technique of choice to delineate the details of pathological anatomy of the malformation especially regarding composite lesions].

b) The neuroimaging findings of limited dorsal myeloschisis follow evolutionary logic. [The neuroimaging findings of limited dorsal myeloschisis follow embyrogenetic logic]

c) The neuroimaging findings do not illustrate the various anatomic forms. [The neuroimaging findings illustrate precisely the various anatomic forms.]

d) The defining feature presents in all limited dorsal myeloschisis is the fibroneural stalk connecting the base of the cutaneous lesions to the underlying spinal cord following a discrete extraspinal and then intrathecal course. (applies)

e) The entire ‘‘tract’’ is usually traceable on CT. [The entire ‘‘tract’’ is usually traceable on MRI.]

Applies to article: *Kameda-Smith M, Tahir MZ, Kumar A, Thompson D, Pang D. Limited exposure to preserve stability and achieve complete excision of limited dorsal myeloschisis - the "Skip-Hop Laminectomy" technique: a technical note. Childs Nerv Syst. 2024 Jan;40(1):213-218. doi: 10.1007/s00381-023-06069-x and Wong ST, Pang D. Focal Spinal Nondisjunction in Primary Neurulation: Limited Dorsal Myeloschisis and Congenital Spinal Dermal Sinus Tract. J Korean Neurosurg Soc. 2021 Mar;64(2):151-188. doi: 10.3340/jkns.2020.0117.*

5 - Which of the following statements is true about limited dorsal myeloschisis?

a) Antenatal diagnosis of limited dorsal myeloschisis can often resemble myelomeningocele.

b) LDMs cause neurological deficits rarely through their tethering effect.

c) The type and severity of neurological deficits do not depend on the spinal level affected by the limited dorsal myeloschisis.

d) The surgical indication of limited dorsal myeloschisis involves the maintenance of the stalk to untether the cord.

e) The surgical indication of a limited dorsal myeloschisis is well doubtful.

Explanation:

a) Antenatal diagnosis of limited dorsal myeloschisis can often resemble myelomeningocele. (applies)

b) LDMs cause neurological deficits rarely through their tethering effect. [Limited dorsal myeloschisis cause neurological deficits exclusively through their tethering effect]

c) The type and severity of neurological deficits do not depend on the spinal level affected by the limited dorsal myeloschisis. [The type and severity of neurological deficits depending on the spinal level affected by the limited dorsal myeloschisis]

d) The surgical indication of a limited dorsal myeloschisis involves the maintenance of the stalk to untether the cord. [The surgical indication of limited dorsal myeloschisis involves relatively straightforward resection of the stalk to untether the cord.]

e) The surgical indication of a limited dorsal myeloschisis is well doubtful. [The surgical indication of a limited dorsal myeloschisis is well established.]

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FIGURES

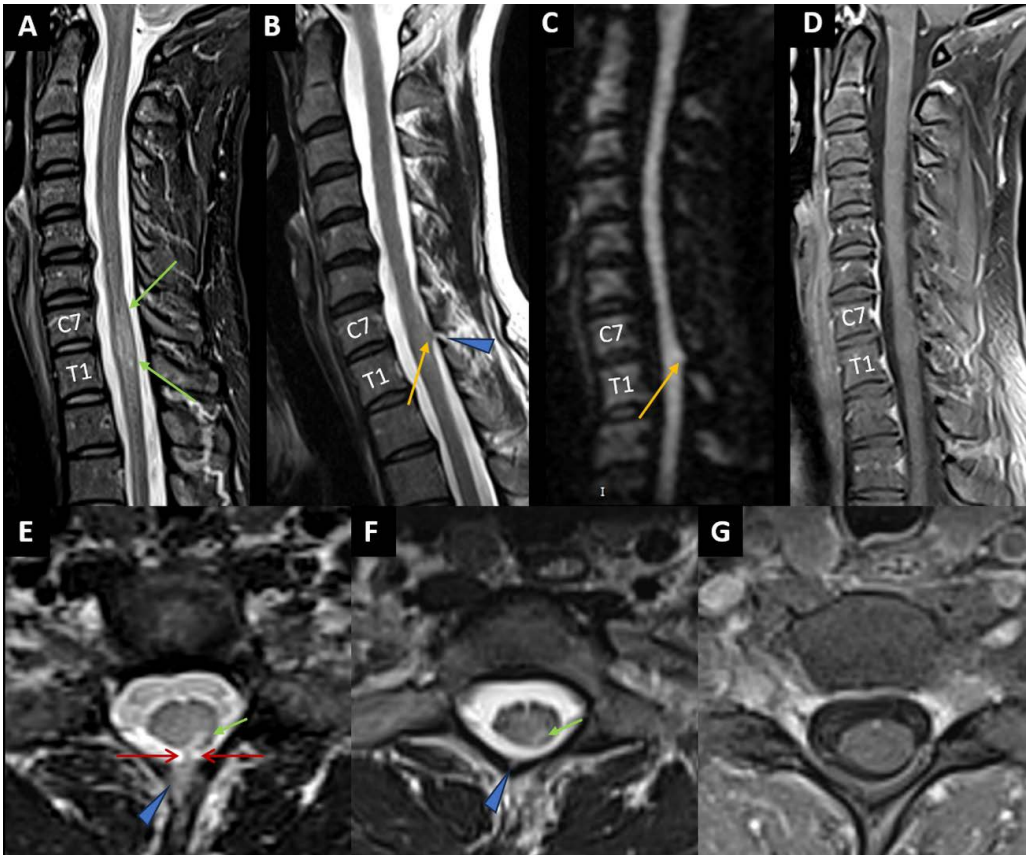

Figure 1: 37-year-old female patient with limited dorsal myeloschisis. Cervical spine MRI on sagittal STIR (A), T2 (B), diffusion (C) and fat-saturated post-contrast T1 (D) weighted images; axial T2 (E-F) and fat-saturated post-contrast T1 (G) weighted images. There was a hyperintense eccentric expansile intramedullary lesion at C7-T1 (green arrows), with an exophytic and triangular shape of the left posterolateral aspect (orange arrows), extending toward the dura mater. Note is made for the focal tiny disruption of the posterior dura mater (blue triangles) and a fibrous tissue (rudimentary neural stalk – red arrows) connecting both. No abnormal enhancement was appreciated (D and G).

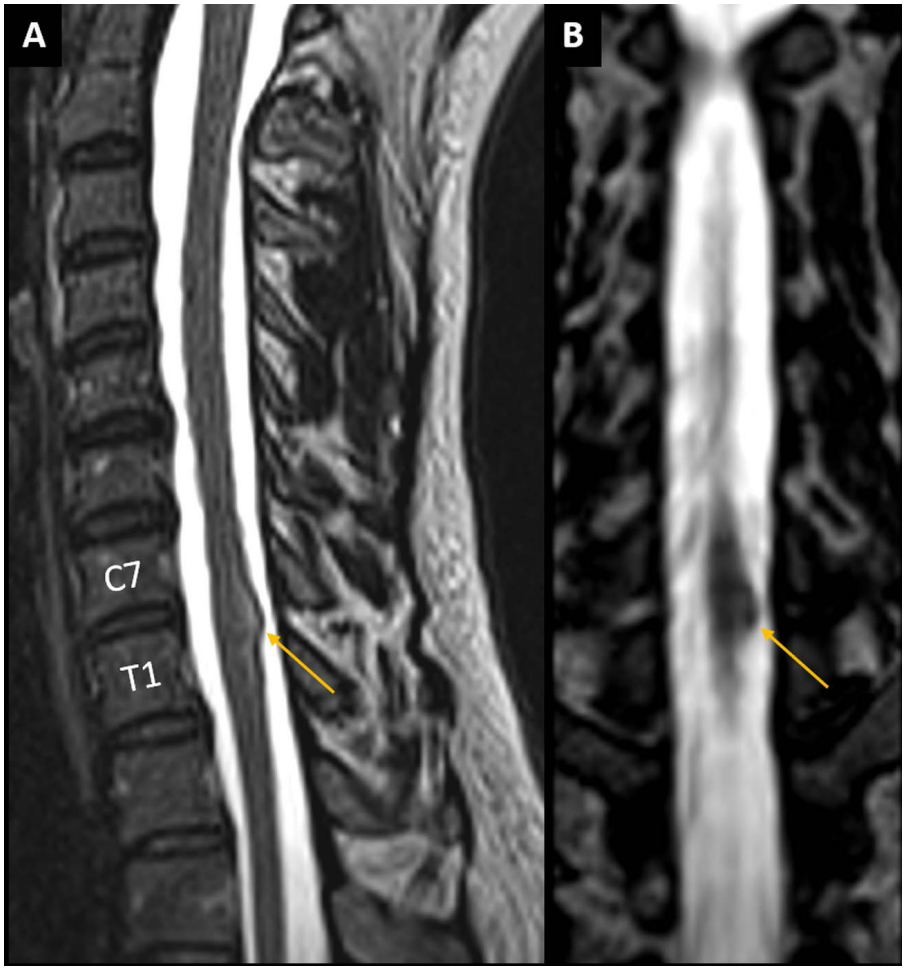

Figure 2: – 37-year-old female patient with limited dorsal myeloschisis. 3D-CISS sequence of cervical spine MRI on sagittal (A) and coronal (B) view, with 0.9 mm slice thickness. Greater conspicuity of the exophytic and triangular aspect ("beaking" – orange arrows) of the left posterolateral C7-T1 spinal cord, extending toward the posterior dura mater.

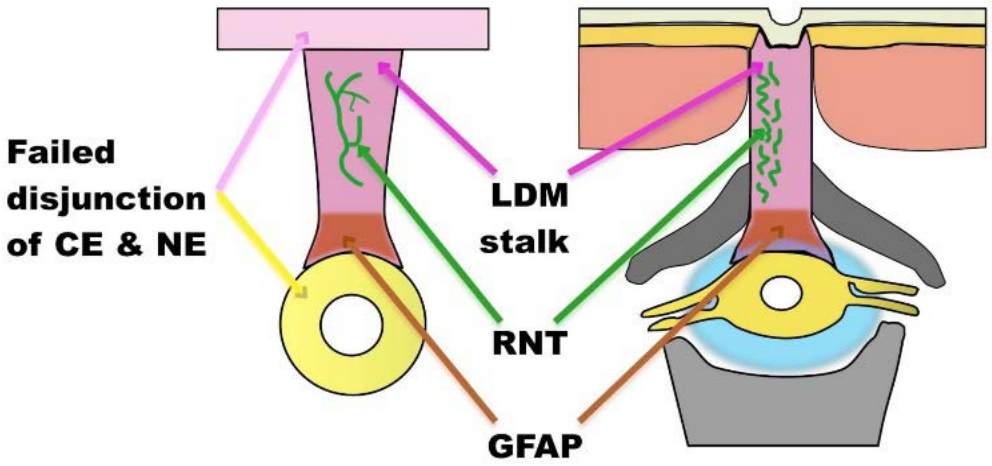

Figure 3: Schematic drawing of the spinal cord (yellow) and its surrounding structures during abnormal primary neurulation. LDM originates from a focal incomplete disjunction between EC and NE, with a persistent fibroneural stalk (pink), characterized by GFAP positivity (brown) and rudimentary neural tissue (RNT - green), between the skin and the dorsal spinal cord, causing tethering of the spinal cord. *Modified from Murakami N et al.[1]*

KEYWORDS

Limited Dorsal Myeloschisis; Magnetic Resonance Imaging; Spinal Cord; Spine; Dysraphism

ABBREVIATIONS

MRI = Magnetic Resonance Imaging
LDM = Limited Dorsal Myeloschisis
CT = Computed Tomography

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