

Midline Spinal Cord Hamartoma Accompanied by the Lipoma of the Filum Terminale

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Abstract

The midline spinal cord hamartoma is a rare non-hamartomatous malformation that is often accompanied by a dermal sinus tract. Lipoma of the filum terminale is one of the subtypes of closed spinal dysraphism and it is characterized by lipomatous tissue that can inclose part or all of the filum terminale. A 5-year-old girl has soft tissue mass and hair growth that was present from birth in the posterior neck. In cervical MR examination, protrusion at the cervicomedullary junction and dermal sinus tract was observed. Also, in the lumbar MR examination, lipoma of the filum terminale has shown. Midline spinal cord hamartoma is an extremely rare malformation and the lipoma of the filum terminale associated with this malformation have not been shown before.

Keywords

Midline spinal cord hamartoma, Lipoma of the filum terminale, Spinal cord malformation, Skin tag, Dermal sinus

Introduction

The midline spinal cord hamartoma is a rare non-hamartomatous malformation that is often accompanied by a dermal sinus tract associated with the skin dimple or soft tissue mass [1]. This malformation is rarely defined in the literature and it is not associated with hamartomatous diseases or brain malformations as opposed to known hamartomas. Patients are usually asymptomatic except for skin lesions. Lipoma to the filum terminale is one of the subtypes of closed spinal dysraphism. It is characterized by lipomatous tissue that can inclose part or all of the filum terminale and may be asymptomatic or cause tethered cord syndrome [2]. In this case report, we aimed to present the midline spinal cord hamartoma and lipoma of the filum terminale who presented with a complaint in the soft tissue in posterior neck, which were first shown in a patient.

Case Report

A 5-year-old girl was brought to the hospital by her family because of the soft tissue mass and hair growth that was present from birth in the posterior neck. On physical examination, a non-specific exophytic soft tissue appearance was observed in the posterior neck of the skin about 1 cm in diameter. The patient's neurological examination was normal and there was no other complaint. In the cervical MR examination, a homogeneous lesion was observed skin in the posterior neck of the patient, 8 x 9 mm in size, continuing with the dermal sinus tract. It was observed that the dermal sinus tract extended to the cervicomedullary junction level and the spinal cord protruded posteriorly in this segment

(Figure 1-3). Brain, thoracic and lumbar MRI examinations were performed in order to search for additional anomalies in the patient. Brain and thoracic MR examinations were evaluated as normal. Lumbar MR examination revealed a hyperintense, lipomatous lesion on T1- and T2-weighted images with a size of 34 x 5 mm at the level of the filum terminale (Figure 4 and 5). The patient was diagnosed with midline spinal cord hamartoma and lipoma of the filum terminale.

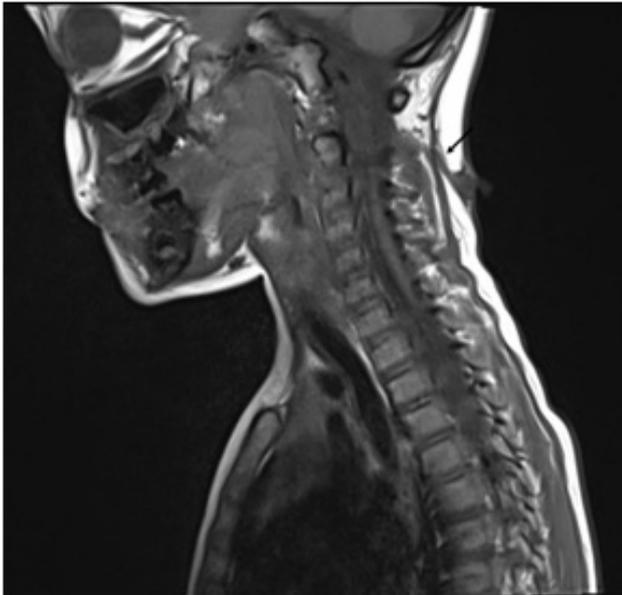


Figure 1: Sagittal T1WI, dermal sinus tract extending from the subcutaneous fat tissue to the spinal canal (black arrow).

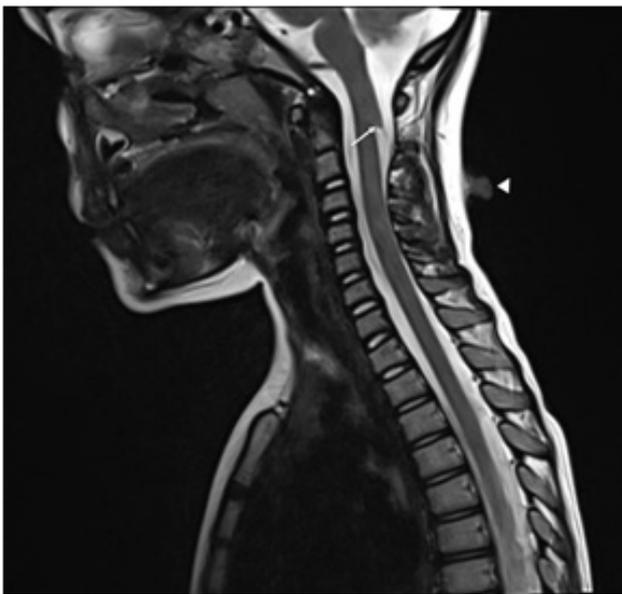


Figure 2: Sagittal T2WI, extension of the dermal sinus tract to the posterior notch to the cervicomedullary junctional level (white arrow). Homogeneous exophytic soft tissue extending from skin in posterior cervical region (white arrowhead).

Discussion

Common congenital lesions of the spinal cord are

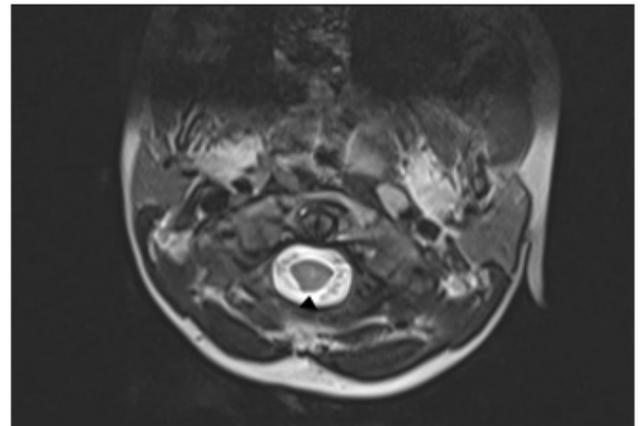


Figure 3: Axial T2WI, posterior notch at cervicomedullary junctional (black arrowhead).



Figure 4: Sagittal T2WI, hyperintense lipoma of the filum terminale (black arrow).

lipomas, epidermoids, teratomas and dermoids [3]. Midline spinal cord hamartoma is a rare malformation associated with dermal sinus tract and skin pathologies [1]. This lesion, which is not associated with cranial anomalies and hamartomatous diseases, is often asymptomatic except for skin pathologies [1]. Mechanism of spinal hamartomas not known. In the embryonic process, the neural tube closes on the 28th day and is wrapped by mesenchyme. If the neural tube cannot be completely separated from the ectoderm layer, the dermal sinus tract is formed between the skin and the spinal cord. Because of this tract, the mesenchyme closes dorsal aspect of the neural tube and results creation of spina bifida. In another hypothesis, the early disjunction between the neural tube and the ectoderm, dorsal mesenchyme, is allowed to enter the neural groove and contact with the internal side of the unfolded neural tube. The neural tube stimulates primordial mesenchymal precursors, which can be differentiation into



Figure 5: Sagittal T1WI, hyperintense lipoma of the filum terminale (black arrow).

other mature mesenchymal elements. Castillo et al. argued that both ways could be effective in the process [1]. In the study of Rao et al., it is stated that the mechanism of formation may be due to incomplete fusion in the neural tube [4]. It has also been described that the protrusion in the spinal cord may be caused by a traction of the dermal sinus tract [1]. In histopathological examination, dense connective tissue, meningoepithelial proliferation and calcification in a study [1]. In the interview with the patient's family, because of the complications of the procedures to be performed had a negative effect on the patient's life, they refused any interventional procedure. We decided to follow the patient without interventional procedure because the family did not want and this malformations does not threaten patient's life. At the differential diagnosis, meningomyelocele may be considered due to polypoid soft tissue present at the skin [5]. However, the absence of defect in bone structures removes meningomyelocele. Epidermoid tumors are distinguished by T1AG hyperintensity. Teratomas, on the contrary to the midline spinal cord hamartoma, are heterogeneous internal structures [1]. Non-symptomatic

patients are followed up, and if there are additional anomalies, a treatment plan should be prepared for them.

One of the closed spinal dysraphism subtypes, the lipomas to the filum terminale can retain the filum terminale as total or segmental. The incidence of this lesion, which is not known precisely in the society, has increased with the increase in the number of diagnostic examinations [2]. Lipomas to the filum terminale are usually asymptomatic but have been reported to cause tethered cord syndrome in some cases [6]. When symptomatic, there may be complaints such as urological complaints, back pain, leg pain, etc. At the differential diagnosis, paraganglioma of the filum terminale and mixopapillary ependymoma should be considered [7]. However, they are both hypointense in T1WI and enhancing postgadolinium that distinguished from lipomas. Non-symptomatic patients are followed up, patients with tethered cord syndrome are treated surgically.

Conflict of Interest

The authors declare no conflict of interest.

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