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Tethered Spinal Cord with Double Spinal Lipomas

Although lumbosacral lipoma is reported to occur in 4-8 of 100,000 patients, and 66% of lipomyelomeningoceles in young patients are accompanied by hypertrophic filum terminale, it is very rare to find two isolated spinal lipomas simultaneously. A 3 month-old baby girl was admitted to the hospital for a protruding, non-tender, soft, subcutaneous 2.5 cm mass of the lumbosacral area that had been present since birth. Simple radiography showed a spinal posterior arch defect from L3 to L5, and magnetic resonance imaging (MRI) demonstrated two isolated spinal lipomas, a transitional type from L3 to L5, and a terminal type below S1 without dural defect. The cornus medullaris was severely tethered descending to the S1, but there was no cerebellar or brain stem herniation on the MRI. We suggest that the presence of a combined spinal lipoma should be a point for careful differentiation in an infant with spinal lipoma.

Key Words : *Lipoma; Spinal Dysraphism; Tethered Cord Syndrome; Neural Tube Defects*

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INTRODUCTION

Lumbosacral lipoma which occurs in 4-8 per 100,000 of the general population is reported as the most common cause of tethered cord syndrome and 1/4,000 new born babies are born with this disease (1, 2). In addition, lipomyelomeningocele (LMM) was reported to be present in 25-30% of children with tethered cord syndrome (3, 4). Although 66% of LMM in young patients are accompanied with hypertrophic filum terminale, it is rare to find two isolated spinal lipomas simultaneously. In this report, we describe a baby girl with two combined spinal lipomas of transitional and terminal types.

CASE REPORT

A 3-month-old female baby presented with a 2.5 cm sized protruding, non-tender, soft, subcutaneous mass in the lower lumbosacral area which was observed since birth. There were no specific abnormal findings on the neurologic examination. However, multiple spinal posterior arch defects in the lower L3-4-5, as well as the sacral area were observed. The magnetic resonance imaging (MRI) demonstrated two large subcutaneous fat masses with high signal intensities in T1- and T2-weighted images from the L3 area to the sacrum (Fig. 1). Through the bony defect from L5 to S1, the subcutaneous fat was connected to the spinal cord that was found as a spinal lipoma of the transitional type in the area of L3 to L5. Additionally, the other spinal lipoma of the terminal type was observed on the filum teminale below the S1 caudally. Both lipomas appeared to be continuous with large subcutaneous fat tissue. There was no evidence of abnormal signal change in the whole spinal cord. Although the cornus medullaris showed severe tethering down to the S1 area, there was no cerebellar or brain stem herniation shown on the MRI.

Surgery was conducted by a skin incision from T12 to the S2 area. Multiple laminectomy of L2 to L5 revealed an intact dura without any direct communication of the spinal lipoma with subcutaneous fat tissue. After dural incision, cord detethering was achieved by removal of two spinal lipomas of the upper transitional type and lower terminal type, which was adhered to the tethered cord in the L3 to L5 and filum terminale in the S1 to S3 area, respectively (Fig. 2). Pathologic examination revealed benign adipose and fibrous tissue without neuroglial element that was consistent with lipoma. The postoperative course was uneventful. She was followed-up to 12 months without any neurologic or urologic abnormalities.

DISCUSSION

Congenital lumbosacral lipoma or LMM is an embryogenic origin disorder of occult spinal dysraphism which makes up the majority of occult spinal dysraphisms, and 1/4,000 new born babies are born with congenital lumbosacral lipoma (1, 2). LMM is described in various terms such as spinal lipoma (5, 6), congenital lumbosacral lipoma (7, 8), and spinal bifida with lipoma (9, 10) in the literature. Thereafter, there emerged some differing opinions that the term of LMM is not correct because of the replacement of neural elements not by herni-

ation of neural tissue but by a lipoma (6). French confined the term lipomeningomyelocele to the protrusion of neural elements from the spinal canal accompanying a dural defect (11). It is preferable to employ the term simple lipoma when there is no accompanying dural defect (7). However, there is as yet no consensus on a clear terminology when dural or bony defects are present (4).

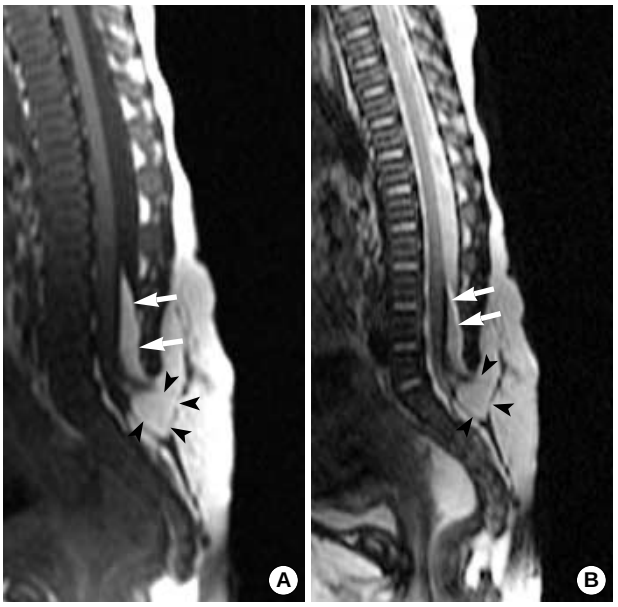

Fig. 1. Initial preoperative spine MR of T1-weighted (A) and T2-weighted images (B) shows tethered cornus medullaris to the level of sacrum and two isolated lipomyelomeningoceles, a transitional type from L3 to L5 (arrows), and a terminal type below S1 (arrow heads).

The most widely accepted classification of spinal lipoma is 3 types; dorsal, transitional, and terminal lipoma, defined with regard to the connection with the cord, conus medullaris, or filum terminale (12, 13). The terminal lipoma is contiguous from the terminal conus replacing the filum terminale (10). It has been previously reported that the terminal type shows good prognosis while the transitional type is very poor (3, 14). However, it was suggested that even the terminal type could result in poor postoperative results when the terminal lipoma is attached to the conus, compared with good results when it is attached to the filum teminale (15). Following this concept, Arai et al. (7) recently re-classified the terminal type to the caudal type and filar type, when the terminal lipoma is attached to the conus and the filum terminale, respectively. They also included a lipomeningomyelocele type as defined by French into their modified classification.

Even though there are many reports with regard to simultaneous occurrences of meningocele (16), myelomeningocele (17, 18), and LMM or spinal lipoma (19-21) in different spinal levels of all spinal dysraphisms, there are very few reports to date regarding the presence of two simultaneous, isolated, non-consecutive spinal lipoma of dorsal and terminal or filar type occurring in the same spine as shown in this report (22).

McLone and Naidich (6) discussed the hypothesis of premature dysjunction of the upper LMM formation which is similar in the dorsal and transitional types of LMMs. In this hypothesis, the folding neuroectoderm leaves a cleft dorsally, allowing the paraaxial mesenchyme access to the prospective lumen of the neural tube. The luminal surface of the neural tube induces the mesenchymal cells to differentiate

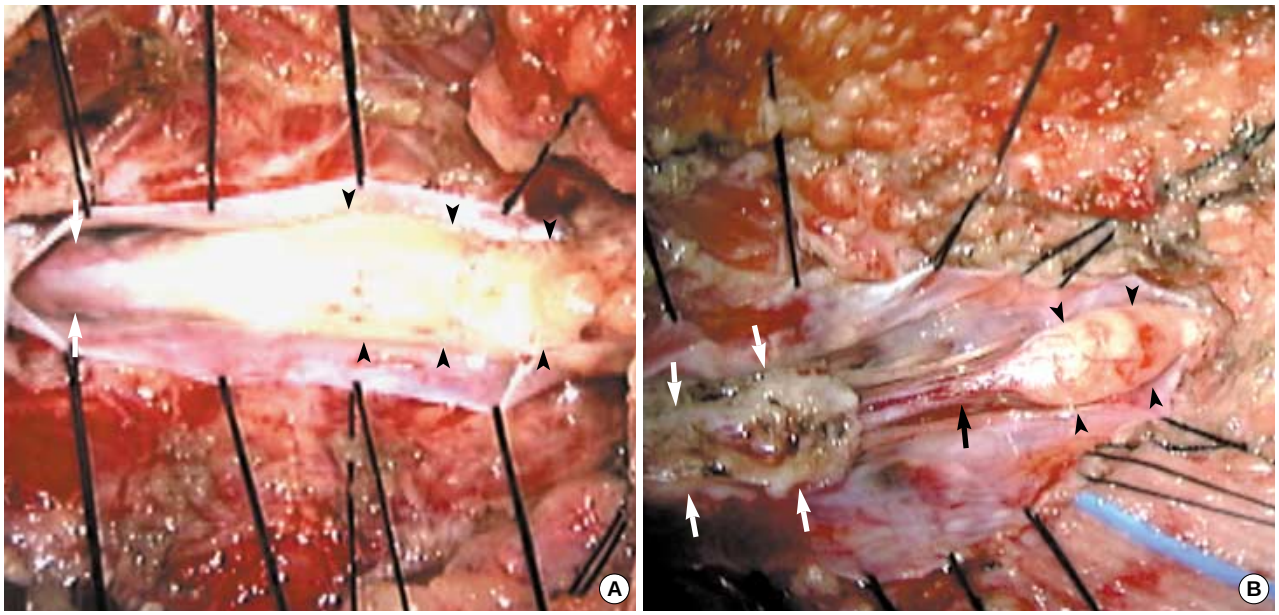

Fig. 2. (A) After incision of the dura of L2 to S1, the normal cord (arrows) and transitional type spinal lipoma (arrow heads) are exposed. (B) Another terminal type spinal lipoma (arrow heads) was exposed and removed from the filum terminale (black arrow) after upper transitional type spinal lipoma was detethered and dissected (white arrows).

along a path, resulting in adipocyte formation. Regarding the period in which the terminal type spinal lipoma of the lower spinal cord occurs by secondary neurulation, it is explained by the maldegeneration hypothesis of the caudal cell mass (23). This theory suggests that this is when adipose tissue expands from the end of the distal part of the spinal cord, and during the process of filum terminale or cornus medullaris replacing adipose tissue. It also suggests that the lipoma around the filum terminale area results from maldevelopment of the tail bud during the period of undifferentiated multipotent cells during secondary neurulation (24). On the other hand, Catala (9) suggested another hypothesis, abnormal development of dorsal mesoderm with multi-potent cells, which overcomes the limitations of the above two theories by pointing out that development of the dorsal mesoderm is regulated by different molecular biologic controls than the rest of the somites. Catala's hypothesis of multi-potent cells was supported clinically by the presence of development of a teratoma from the spinal lipoma (25). Classically, we expect that both the premature dysjunction mechanism and maldegeneration processes of the caudal cell mass in secondary neurulation were coincidentally involved in different embryonic periods, resulting in two different isolated types of spinal lipoma (24).

We present a rare case of two isolated transitional and terminal type spinal lipomas appearing simultaneously. We suggest that the presence of combined spinal lipomas should be a point for careful differentiation in an infant with spinal lipoma.

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