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 newborn 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. 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.

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).

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 ter-
nimal lipoma is attached to the conus, compared with good
results when it is attached to the filum terminale (15). Follow-
ing this concept, Arai et al. (7) recently re-classified the ter-
nimal type to the caudal type and filar type, when the termi-
nal 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 simul-
taneous occurrences of meningocele (16), myelomeningocele
(17, 18), and LMM or spinal lipoma (19-21) in different spi-
nal 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 pre-
mature 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 paraxial mesenchyme access to the prospec-
tive lumen of the neural tube. The luminal surface of the
neural tube induces the mesenchymal cells to differentiate

Fig. 1. Initial preoperative spine MR of T1-weighted (A) and T2-
weighted images (B) shows tethered conus 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).

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 tran-
sitional 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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