Double Lumbosacral Lipomas of the Dorsal and Filar Types Associated With OEIS Complex

—Case Report—

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Abstract

A female baby was born at 37 weeks and 6 days gestation by vaginal delivery with omphalocele, exstrophy of the cloaca, and imperforate anus, indicating the presence of OEIS complex, a rare combination of defects consisting of omphalocele (O), exstrophy of the cloaca (E), imperforate anus (I), and spinal deformity (S), associated with lumbosacral lipoma. The most common associated spinal deformity is terminal myelocystocele, and spinal lipoma is rare. Constructive interference in steady-state magnetic resonance imaging clearly revealed double lipomas, a dorsal-type lipoma, located dorsal to the low-lying conus medullaris, and a filar-type lipoma, revealed by a thickened and fatty filum terminale. After recovery from abdominogenital repairs, debulking of the dorsal-type lipoma and untethering of the spinal cord by sectioning of the filar-type lipoma were performed at the age of 14 months. Neurosurgical treatment for occult spinal dysraphism should be undertaken after recovery from the initial series of major abdominogenital procedures.

Key words: OEIS complex, lumbosacral lipoma, double lipomas

Introduction

OEIS complex is a rare congenital condition consisting of omphalocele (O), exstrophy of the cloaca (E), imperforate anus (I), and spinal deformity (S). Omphalocele, exstrophy of the cloaca, and imperforate anus are all life-threatening conditions, and are treated by pediatric surgeons on a priority basis. In contrast, the most common spinal deformity associated with OEIS complex is occult spinal dysraphism, which is covered with normal skin,
and neurosurgical intervention is deferred until recuperation from the abdominogenital repair.3,4,14,15,18) The most common spinal dysraphism in patients with OEIS complex is terminal myelocystocele,3,7,11,15,18) whereas spinal lipoma is rare.18)

We treated a case of lumbosacral lipoma with tethered spinal cord associated with OEIS complex. The associated lipomas were rare double lipomas of the dorsal and filar types.5,6,13)

Case Report

A female baby weighing 2382 g was born at 37 weeks and 6 days gestation by vaginal delivery. She had omphalocele, exstrophy of the cloaca, and imperforate anus (Fig. 1). Primary closure of the abdominal wall, closure of the urinary bladder, and ileostomy were performed by pediatric surgeons on day 1 after birth. No abnormality on her back was visible, but computed tomography and magnetic resonance (MR) imaging revealed spina bifida occulta below the L5 level and lumbosacral lipoma. Neurosurgical procedures for the spinal dysraphism were deferred in the absence of any definite abnormal neurological finding.

She was admitted to our department after recovery from her abdominogenital problems at the age of 14 months. She had a body weight of 7660 g and height of 68 cm. Neurological examination found very mild weakness of the left ankle and hyperreflexia of the bilateral lower limbs. Sagittal T1-weighted MR imaging demonstrated that the conus medullaris was located at the L3–L4 levels and the lipoma from the L3 area to the sacrum (Fig. 2A). Three-dimensional (3-D) constructive interference in steady-state (CISS) imaging, obtained as described previously,8,10,16) clearly revealed two separate lipomas, a dorsal-type lipoma, situated dorsal to the low-lying conus medullaris (Fig. 2B), and a filar-type lipoma, revealed by a thickened and fatty filum terminale (Fig. 2C), according to Arai’s classification.1) Serial axial T1-weighted MR imaging detected no continuity between these two lipomas (Fig. 2D–F). Cranial MR imaging found no Chiari malformation or hydrocephalus.

Surgery was conducted by laminoplastic laminotomy from the L2 to L5 levels. Most of the dorsal-type lipoma had adhered to the low-set conus. The most caudal part of this lipoma protruded into the subarachnoid space and was easily separated from the fatty filum terminale and cauda equina (Fig. 3A). First, the dorsal-type lipoma was debulked and the conus was reconstructed with interrupted pia-arachnoidal sutures to reduce the surface area available for scarring and subsequent retethering. Then, untethering of the cord was achieved by sectioning the filar-type lipoma, after confirming no muscle response of the lower limbs and anus to electrical stimulation (Fig. 3B, C).

The postoperative course was uneventful and no neurological deterioration was present. Histological examination of the sectioned filum terminale showed embedded clusters of mature adipose cells, consistent with filar-type lipoma.

Discussion

The coexistence of occult spinal dysraphism with cloacal extrophy, as seen in OEIS complex, is well known.4) The

Fig. 1 Photograph of the patient at birth showing the typical findings of omphalocele (arrow), exstrophy of the cloaca (arrowhead), and imperforate anus.

Fig. 2 A: Sagittal T1-weighted magnetic resonance (MR) image demonstrating conus medullaris located at the L3–L4 levels and lipoma from the L3 area to the sacrum. B, C: Sagittal three-dimensional constructive interference in steady-state images clearly revealing two separate lipomas, a dorsal-type lipoma, situated dorsal to the low-lying conus medullaris (B, arrow), and a filar-type lipoma, revealed by a thickened and fatty filum terminale (C, arrow). D–F: Serial axial T1-weighted MR images demonstrating no continuity between the double lipomas (D and F, arrow). Both the dorsal-type lipoma (D) and the filar-type lipoma (F) appear as high signal intensity; but without a high intensity area between the double lipomas (E). Slice levels are indicated on A.
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Fig. 3 A: Intraoperative photograph after laminoplasty of L2 to L5 and dural opening. Most of the dorsal-type lipoma has adhered to the low-set cord. The rostral pole of the dorsal-type lipoma was easily elevated with forceps (arrow). B: Intraoperative photograph showing the filar-type lipoma was elevated with a rubber sheet (arrow) and the cauda equina was exposed. C: Intraoperative photograph showing the dorsal-type lipoma was debulked and the conus was reconstructed with interrupted pia-arachnoidal sutures (arrows). Untethering of the cord was achieved by sectioning of the filar-type lipoma. Thick arrow indicates the cut end of the filar-type lipoma.

Our patient had spinal lipoma instead of terminal myelocystocele. Only one patient with OEIS complex and spinal lipoma of a transitional type has been reported.18 Furthermore, our patient had two different types of lipoma, or “double lipomas.” Two isolated spinal lipomas found simultaneously are rare.6,13 One patient had double lipomas at the L4 and S2 levels, and cloacal exstrophy, but not OEIS complex,5 although we think that the pathophysiology was quite similar to that of our case.

In the present case, sagittal T1-weighted MR imaging failed to reveal the double lipomas, whereas 3-D CISS imaging clearly demonstrated the detailed anatomical relationship between the double lipomas and the spinal cord. We previously reported on the usefulness of 3-D CISS imaging for the preoperative examination of spinal dysraphism.8,10,16 Conventional MR imaging provides sections that are 2–5 mm thick, which is inadequate for detailed imaging of the spinal cord or nerves in infants.8,10,16 3-D CISS images can be reconstructed with a minimum slice thickness of 0.2–0.3 mm in any specified linear or curved plane.5,8,10,16 However, conventional T1-weighted MR imaging is still an excellent modality for the diagnosis of spinal lipoma, since these lipomas appear as high signal intensity. The detailed anatomical relationship between the complicated pathologies can be further delineated by 3-D CISS imaging, which can also serve as a surgical road map.15,17 as in our case.

Whether the associated spinal dysraphism is terminal myelocystocele or spinal lipoma with tethered cord, neurosurgical treatment for occult spinal dysraphism should clearly be undertaken after the infant recovers from the initial series of major abdominogenital procedures.4,15,18 However, there are no established guidelines regarding the optimal timing of neurosurgery.10 Neurosurgical correction of the myelocystocele in seven patients with OEIS complex was undertaken between the ages of 1 and 19 month, with a mean age of 5 months, but there was no theoretical basis regarding the timing of neurosurgery.3 In our previous report, neurosurgical procedures were performed in three patients with OEIS complex weighing 3–4 kg and/or at an age of 3 months, since all patients presented with progressively increasing lumbosacral cystic masses.15 In the present case, serial physical examination and MR imaging showed that the lumbosacral lesion had not changed, so debulking of the lipoma and untethering of the spinal cord were performed at the age of 14 months after recovery from her abdominogenital problems.

References


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