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Case Report

Spinal Extradural Meningeal Cyst in Klippel-Trenaunay Syndrome

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A case of a symptomatic spinal extradural meningeal cyst (SEMC) in Klippel-Trenaunay syndrome (KTS) is introduced. A 38-year-old woman presented with right L2 radiculopathy. She underwent operations for varicose veins in both her lower extremities. She had port-wine nevi on her trunk and extremities. The edematous change in both legs had waxed and waned. Magnetic resonance imaging showed an 11.8×13 mm extradural meningeal cyst growing through the intervertebral foramen in L2-3. Multiple meningeal cysts were located in the dorsal aspect of the spinal cord from T3 to T10. A 5.8×6.2 mm cyst was also found in left pleural cavity. The extradural meningeal cyst was completely excised and the preoperative symptom was improved. KTS is a congenital disorder due to a mesodermal abnormality, which may predispose the dura to weakness. The SEMC may occur through the dural defect or weakened point.

Key Words : Spinal extradural meningeal cyst · Klippel-Trenaunay syndrome · Mesodermal abnormality.

INTRODUCTION

A spinal extradural meningeal cyst (SEMC) is a rare disease and a cause of neural compression. SEMC, first introduced in 1934[10], has been described as a diverticulum of the dura, pouch, and arachnoid cyst[18,20,24].

Klippel-Trenaunay Syndrome (KTS) is a rare congenital syndrome characterized by a triad of port-wine stains, varicose veins, and a bony or soft tissue hypertrophy involving an extremity.

We present a case of radiculopathy caused by extradural meningeal cyst in Klippel-Trenaunay syndrome.

CASE REPORT

A 38-year-old woman presented with right buttock and lateral thigh pain. Upon admission, she had difficulty in standing and walking due to severe pain. She was unable to sleep due to pain which did not respond to analgesics. The patient had undergone operations for the removal of soft masses of the knee and ankle twice in orthopedics. She also underwent operations for varicose veins in both lower extremities. She had port-wine nevi on her trunk and extremities. The edematous change of both her legs had waxed and waned (Fig. 1). A plain lateral radiography revealed widening of the L2-3 intervertebral foramen. Magnetic resonance imaging (MRI) showed an 11.8×13 mm extradural cyst that contained cerebrospinal fluid (CSF) intensity collections growing through the intervertebral fora-

Fig. 1. Photograph shows edematous right leg and multiple port-wine stains.

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Fig. 2. T2-weighted axial image shows a large extradural cystic mass in the intervertebral foramen of L2-3.

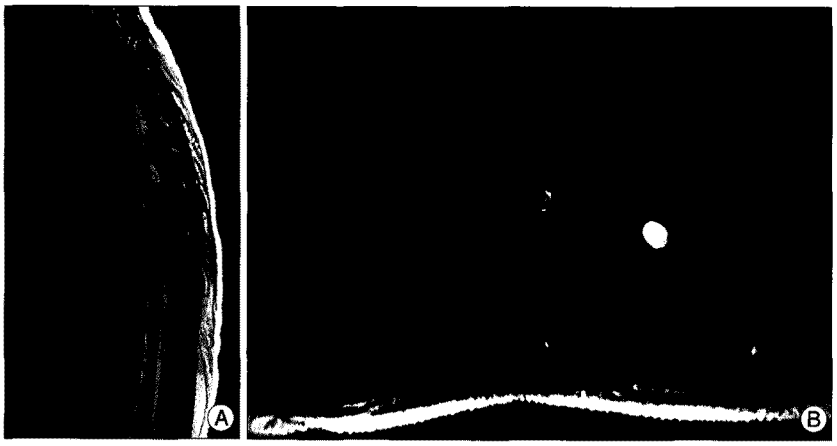

Fig. 3. Thoracic T2-weighted sagittal image (A) shows multiple extradural cysts in the entire thoracic spine and T2-weighted axial image (B) shows a cyst in the left pleura.

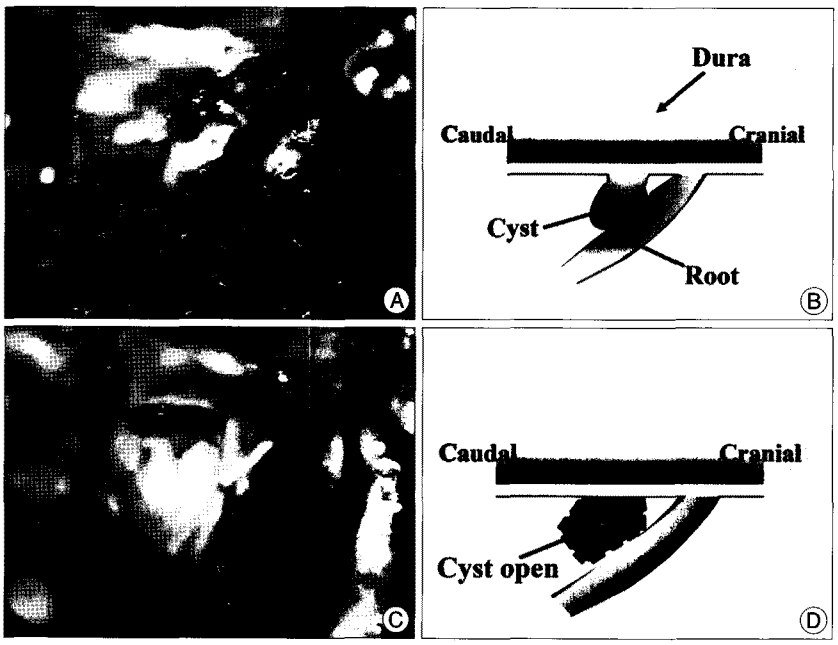

Fig. 4. A : Intraoperative photograph showing the extradural cyst which is located at adjacent nerve root compressing the nerve root. B : Schematic illustration of the extradural meningeal cyst in A and C : After the opening of the cyst, no nerve fiber is detected. D : Schematic illustration of opening the cyst in C.

men in L2-3 (Fig. 2).

Thoracic MRI demonstrated multiple dorsal epidural CSF intensity masses with a mild mass effect from T3 to T10 (Fig. 3A). A 5.8×6.2 mm cyst was also found in left pleural cavity (Fig. 3B).

The meningeal cyst was removed using a lateral transmuscular approach. The cyst was located just below the pedicle. The cyst, which was compressing the right L2 exiting nerve root, was not connected with the nerve root, although there was a minimal adhesion between the cyst and L2 nerve root (Fig. 4A, B). The wall of the cyst was incised and nerve fiber was not observed (Fig. 4C, D). The cyst was completely removed and the ostium of the cyst was ligated with sutures. After surgery, the patient's leg pain was significantly improved.

DISCUSSION

Spinal extradural meningeal cyst (SEMC) is a rare cause of radicular pain which accounts for approximately 1 to 3% of all primary space-occupying lesions[10,17]. Nabor et al.[19] classified spinal meningeal cysts into three categories : extradural meningeal cysts without nerve root fibers (Type I), extradural meningeal cysts with nerve root fibers (Type II), and intradural meningeal cysts (Type III). The spinal extradural meningeal cyst is involved most commonly in the thoracic spine followed by the lumbar, lumbaosacral and thoracolumbar spine[6].

Various etiological possibilities for spinal extradural meningeal cysts have been discussed and a congenital defect is one of causes[4]. SEMC is also caused by inflammation, trauma, or iatrogenic factors that induced a dural weak spot[24]. These result in herniation of the arachnoid through a weak spot in the dura. The development of Type I SEMC may be attributed to the congenital diverticulum of the dura or arachnoidal herniation due to a congenital dural defect. This is supported by hereditary syndrome[4,7,26], familial tendency[4], or associated congenital anomalies[11,22]. The hereditary syndrome of multiple congenital extradural cysts is associated with distichiasis and lymphedema[14]. There is an alteration of the FOXC2 gene with this syndrome. The FOXC2 gene is expressed in the developing mesodermal mesenchyme of the head, kidney, and bones. It is also expressed in the developing heart, vessels,

and limbs and is essential for normal development of the aortic arch and axial skeleton. KTS consists of a triad of cutaneous capillary hemangiomas, bone and soft tissue hypertrophy, and venous varicosities and these triad was present in the present case[13,16,21]. The manifestations of KTS are variable[21]. The venous abnormalities usually involve the affected extremity and present as superficial varicose veins. In addition, many patients have an abnormality of the deep venous system of the extremity. Lymphatic abnormalities, cutaneous capillary hemangioma, and a swollen or circumferentially enlarged extremity are manifested. Some patients have macrodactly and a localized mass on the back, chest, or entire extremity. KTS also may involve the central nervous system, such as, macrocephaly, arteriovenous malformations, and intraspinal angiomas[1,8,25].

KTS is caused by mesodermal abnormalities during fetal development[2]. A mesodermal defect, acting primarily on angiogenesis could explain vascular malformations and result in bone and soft tissue abnormalities[27]. Mesodermal abnormality may be the causative factor for a congenital dural defect. SEMC in KTS may occur through a dural defect at the root sleeve junction with the dural sac or a weak spot in the dura by remnant arachnoid cells[9,11].

The growth and enlargement of the SEMC is not fully understood. Many mechanisms were explained for its growth. The existence of an osmotic gradient between the subarachnoid space and cyst may expand the SEMC[12]. Hydrostatic pressure of CSF and ball-valve mechanism promote enlargement of the cyst[18]. Some authors proposed that active fluid secretion of the cell wall expands the cyst[11,20].

MRI is helpful in demonstrating an extradural cystic structure with CSF signal intensity. MRI can also help in identifying displacement of the epidural fat and subarachnoid space, inclusion of nerve rootlets and extension into intervertebral foramina.

Surgery is usually recommended in symptomatic SEMC. The cyst should be completely resected and the dural defect should be closed. Surgically treated SEMC has a 75% good outcome rate[23]. The outcome depends on the age, the duration, and degree of neurological deficit[2,15].

CONCLUSION

The SEMC associated with KTS may be caused by mesodermal abnormality.

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