

Symptomatic Foraminal Extradural Meningeal Cyst

Semptomatik Foraminal Ekstradural Meningial Kist

ABSTRACT

The authors described the case of a 39-year-old man with Klippel-Trénaunay syndrome, who had an extradural meningeal cyst expanding into intervertebral foramen of lumbar 2 and 3 vertebrae. The patient suffered from low back pain radiating to the left lower extremity. Magnetic resonance imaging revealed a huge extradural meningeal cyst growing through intervertebral foramen far laterally. A widened neural foramen of L2 and L3 vertebrae was observed on his plain radiography. The cyst was totally excised after tying the ostium connecting the subarachnoid space of the dural sac. This case supports the congenital theory in the pathogenesis of spinal cysts because the Klippel-Trénaunay syndrome is a congenital disorder including a mesodermal abnormality which may be the causative factor for a congenital defect in dura.

KEYWORDS: Cyst, Extradural, Neural foramen, Klippel- Trénaunay syndrome, Meningeal, Spinal

ÖZ

Yazarlar, Klippel- Trénaunay sendromu olan 39 yaşındaki erkek bir hastada lomber 2-3 seviyesinde intervertebral forameni genişleten bir extradural meningeal kist oluşumunu sundular. Hastanın sol bacağına yayılan bel ağrısı yakınması vardı. Manyetik rezonans görüntüleme, intervertebral foramenin far lateral büyüyen büyük meningeal kist varlığını gösterdi. Düz radyografisinde L2-L3 omurgalarının oluşturduğu intervertebral foramenin genişlemiş olduğu görüldü. Dural kesenin subaraknoid aralığı ile bağlantıyı sağlayan açıklığın kapatılmasından sonra kistin tamamı çıkartıldı. Hasta, mezodermal gelişim anomalisinin eşlik ettiği Klippel-Trénaunay sendromuna sahip olduğundan omurga kistlerinin patogenezinde duradaki gelişim defektinin etken bir faktör olabileceğini öneren konjenital teoriyi desteklemektedir.

ANAHTAR SÖZCÜKLER: Ekstradural, Kist, Klippel- Trénaunay sendromu, Meningial, Neural Foraman, Spinal

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INTRODUCTION

Spinal extradural meningeal cysts are uncommon and rarely cause neural compression. Previous investigators have called extradural meningeal cysts as extradural cysts (2,4), pouches (14) and diverticula (6). Together with other spinal cysts including perineurial root cysts (22), they are indistinct and confusing. Therefore, in order to simplify their description, Nabors et al (15) proposed a classification of spinal meningeal cysts into three major categories: extradural cysts without nerve root fibers (Type I); extradural cysts with nerve root fibers (Type II); and intradural cysts (Type III).

The Klippel- Trénaunay syndrome is a congenital disorder characterized by the triad of cutaneous vascular nevi, soft tissue or bony hypertrophy, and varicose veins or venous malformations involving one or more extremities (11). We have described a patient with such syndrome that develops Type I spinal extradural cyst expanding neural foramen at the levels of L2-L3 and causing L3 radiculopathy.

CASE REPORT

A 39-year-old male patient with Klippel-Trénaunay syndrome was referred to our clinic due to a complaint of low back pain radiating to the left lower limb for the last two months. His physical examination revealed many surgical scars on the hypertrophic and edematous (lymphedema) left lower extremity due to multiple operations for varicose veins and deep venous thrombosis (Figure 1). He also had port-wine stains and widened varicose veins on his body and lower extremity (Figure 2A,B). The neurological examination showed 25-30% strength loss in knee extension and a 15-20% strength loss in dorsal flexion of the foot. The Achilles tendon reflex was (++) , while the patella tendon reflex was (+) on the left leg. Plain lumbar radiography revealed a widened intervertebral foramen of L2 and L3 vertebrae due to erosion of bony tissue on the left side (Figure 3). Magnetic resonance imaging (MRI) of the lumbo-sacral spine showed a huge cystic lesion arising from the spinal canal and expanding into the neural foramen of L2-L3 vertebrae on the left side (Figure 4A,B). This lesion could be seen as a mass with low intensity on T1-weighted images and with high intensity on T2-weighted images on MRI, similar to the CSF.



Figure 1. Photomicrograph shows hypertrophic and edematous (lymphedema) the left lower extremity.

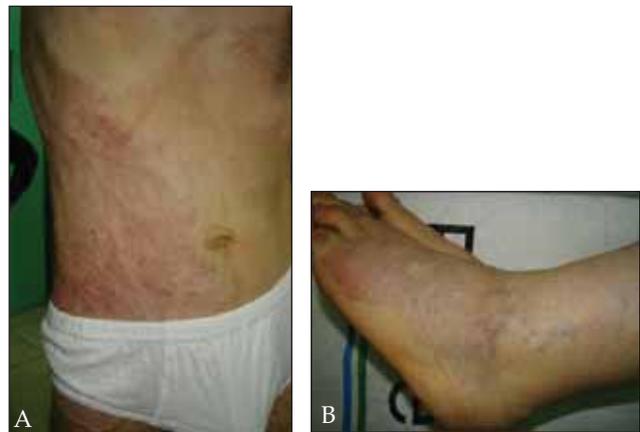


Figure 2. Photomicrographs show extensive Port-wine stains and large sub-cutaneous veins on his body (A), and varicose veins in the left foot (B).

Operation: After partial L2 and entire L3 hemilaminectomy on the left side of spine, we encountered a huge extradural meningeal cyst entering into the intervertebral foramen of L3-L4 levels. There were the dilated epidural veins of Batson's plexus which caused bleeding during the surgery. The cyst compressed the adjacent nerve root within the intervertebral foramen and there was no connection between them (Figure 5A) The extradural meningeal sac had a narrow pedicle near or below of the entrance of the nerve root of L3 (Figure 5B). After dissection of the cyst entirely, the ostium of cyst within the narrow pedicle was ligated with silk

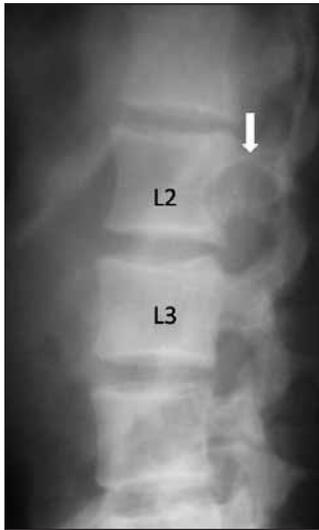


Figure 3. A widened intervertebral foramen of L2 and L3 vertebrae was seen on plain radiography on the left side (Arrow). Note that the bony erosion was not only on pedicle but also on the back of vertebral body of L2.

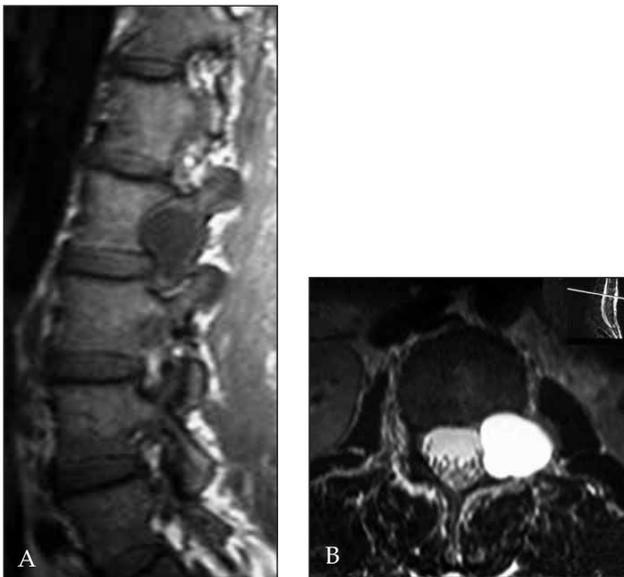


Figure 4. Magnetic resonance image shows a huge cyst expanding through the neural foramen far laterally at the levels of L2-L3 with low intensity on T1-weighted images (A) and with the high intensity on T2-weighted image (B).

sutures. The cyst was incised and no spinal nerve fibers were observed within cyst whereas it contained only cerebrospinal fluid (Figure 5C). Finally, the extradural meningeal cyst was totally excised (Figure 5D). Histopathological examination of the cyst wall showed a delicate fibrocollagenous tissue lined by a single-cell layer of inner arachnoid. The arachnoidal cells stained with epithelial membrane antigen. There was no sign of

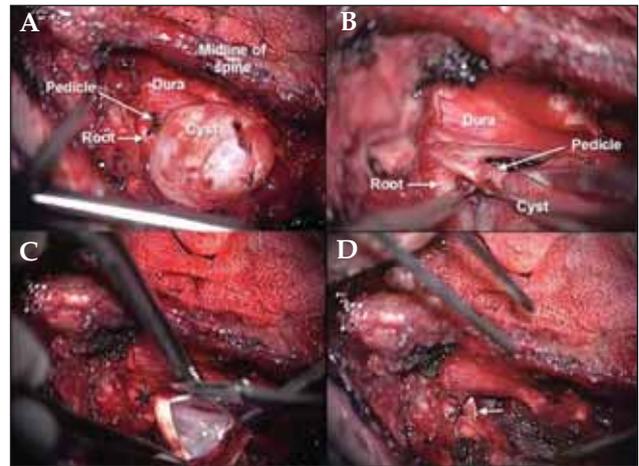


Figure 5. Intra-operative views. Panel A shows the general appearance of the cyst: L2 partial and L3 total laminectomy was performed on the left side. Ostium (pedicle) was closed the silk suture after dissection of the cyst entirely. Panel B shows the localization of pedicle in accordance with the position of L3 root and dural sac. Note that pedicle of the cyst was arisen near or axilla of L3 root. The cyst incised and no spinal nerve fiber was observed (Panel C). The cyst was removed totally after ligation of pedicle (Panel D, arrow).

hemorrhage. The severity of low back pain decreased and radicular pain disappeared after surgery.

DISCUSSION

An extradural meningeal cyst is considered as a "congenital diverticulum of the dura", or a protrusion of the arachnoid through a congenital defect or weakness in the dura (5,22). They are characterized histopathologically by a fibrocollagenous layer with or without a single-cell layer inner arachnoid lining (5,6,15,22). According to Nabors' classification (15), the huge cyst in the present case refers to a Type I lesion, because it has a free communication with the spinal subarachnoid space through the narrow pedicle and there was no connection between the meningeal cyst and the L3 root. The cyst contained no nerve fascicle. A similar observation was reported by others: extradural meningeal cysts are located mostly at the termination of the dural sleeve of the nerve root at its exit from the intervertebral foramen or at the dorsal midline of dura and rarely near or at the junction of the root sleeve and dural sac (3,4,7,9,15,19,22).

The lower half of the thoracic column is involved most frequently, the upper half of the thoracic region and the lumbar region fairly commonly, while the disease is rare in the cervical and sacral regions

(4,19). Clinical features result from nerve root or spinal cord compression and vary with the location of the cyst. Lumbar Type I cysts may present with low back pain and radiculopathy (4,15).

Various etiologies are responsible for development of extradural meningeal cysts. The lesion may be part of a hereditary syndrome or associated with congenital anomalies (12,13,20,24) Congenital extradural cysts are seen associated with the distichiasis and lymphedema syndrome, which includes a double row of eyelashes (distichiasis) and lymphedema in lower extremities (12,24). Hereditary disorders of connective tissue associated with spinal meningeal cysts are Ehlers-Danlos and Marfan's syndromes (10,18). A high incidence of dural ectasia has been noted in patients with Marfan's syndrome (17). An incomplete midline fusion of the embryonic mesenchymal structures surrounding the neural tube, sometimes referred to as dysraphism, may leave a weakened dura and predispose to future cyst formation (7). The Klippel-Trenaunay syndrome is a congenital disorder due to generalized mesodermal abnormality occurring during fetal development (1). This syndrome is associated with a wide spectrum of congenital anomalies including cerebral and cerebellar hemihypertrophy, polydactyly, duplications of sheaths of the posterior roots and macrocephaly (11,16,21,23). Bleeding or deep venous thrombosis are also common features of these syndromes (11). In the present case, there was no evidence of previous hemorrhage and hemosiderin-laden cells in the cyst. The congenital theory including developmental anomaly in the mesoderm layer may explain the development of the spinal extradural meningeal cyst in our patient.

Factors which may cause enlargement of the cyst are 1) the hydrostatic pressure of the cerebrospinal fluid, 2) osmosis of water into the cyst, and 3) secretion by the cyst wall (4,7). Furthermore, some investigators believe that an originally small diverticulum grows by the ball-valve mechanism with the change in spinal pressure associated with the pulse, breathing, and coughing (6,14,19). Bone erosion of the spinal canal may imply the presence of a valve mechanism that is responsible for producing forces of CSF pressure within the cyst that are greater than normal hydrostatic forces (15). Eroded pedicles, widened interpedicular spaces, or scalloped vertebral bodies were frequently seen in

the radiographs of spine in patients with a spinal extradural arachnoid cyst (4,7,22). The enlargement of intervertebral foramina seen on plain lumbar radiography in the present case is due to bony erosion of pedicles of L2-L3 vertebrae possibly caused by pulsatile forces of CSF in cyst.

Surgery is usually recommended in cases involving a large cyst with a mass effect and associated symptoms (8). The total excision of cyst after ligation of the ostium connecting the dural sac is an appropriate option for the treatment such cases (15).

In conclusion, hereditary disorders involving the mesodermal layer during fetal development including the Klippel- Trénaunay syndrome may lead to a predisposition for spinal extradural meningeal cysts.

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