

Case Report

Two Cases of Symptomatic Perineural Cysts (Tarlov Cysts) in One Family - A Case Report

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Symptomatic sacral perineural cysts are uncommon. Several hypotheses have been proposed to explain the etiologies of perineural cysts, but the accurate etiologies remain unclear. We experienced two cases of symptomatic sacral perineural cysts (Tarlov cysts) in one family, who presented with perianal paresthesia. Both of them were operated and postoperatively their symptoms were disappeared immediately. We experienced the excellent treatment outcome with the surgical management of symptomatic perineural cysts in the sacral region. We assume that the theory of congenital origin including a familial tendency is the most plausible of the hypotheses that have been proposed.

KEY WORDS : Tarlov cyst · Sacrum · Congenital.

INTRODUCTION

In 1938, five cases of perineural cysts (Tarlov cysts) in the sacral region were first reported by Tarlov in autopsy findings¹⁵. Although unclear, the etiologies of this disease are assumed to include one of the hypotheses such as inflammation, arachnoid proliferation, trauma, and developmental or congenital origin^{9,12,14,16}. In a series of 500 consecutive magnetic resonance imaging (MRI) scans of the lumbosacral spine, Paulsen et al. recorded an incidence of 4.6%, but the size of cyst has been known to increase and then to cause the symptoms in approximately 1%^{5,11}. To date, only five cases have been reported in Korea^{2,4,6,7}. To our knowledge this is the first report presenting symptomatic sacral Tarlov cyst in one family, and experienced the definite improvement of symptoms following the surgery. Here, we report our cases with a review of literatures.

CASE REPORT

Case 1

This 47-year-old woman was presented with a 20-year

history of right side buttock and perianal pain which were aggravated on standing, sitting and coughing, and relieved by lying down. Two weeks before outpatient visit, the symptoms were worsened. Severe radiating pain was felt to the posterior aspect of her right thigh, which was not alleviated with the further conservative treatments. She had no previous history of trauma, surgery and meningitis. A preoperative hematologic studies showed no abnormal findings. On neurological examinations, she complained of paresthesia in the right S1 and S2 dermatome. But, her motor weakness, bladder and bowel function were all intact.

On lumbo-sacral contrast enhanced MRI scans, there was a cystic lesion in right neural foramen of S2 of 25 × 15 × 25 mm in size whose intensity was the same as cerebrospinal fluid (CSF). No contrast-enhanced findings were observed (Fig. 1A). On initial and a 6-hour delayed myelo-computed tomography (CT) scans, there was no free communication between cyst and subarachnoid space. However, there was the surrounding sacral bony erosion around this cyst (Fig. 2A, B).

A skin incision was made from the L5 to the S3 in the supine position. The adjacent muscles and soft tissue were dissected. The eroded sacral lamina due to the cyst was observed. A laminectomy of the first and second sacral vertebrae was carefully performed. The presence of a translucent, light-brown cyst was identified in the right second lumbar vertebral area. An incision was made on the

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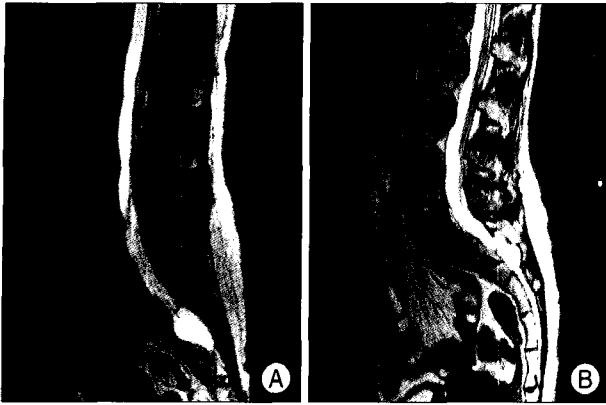


Fig. 1. Preoperative magnetic resonance image (MRI). Preoperative T2-weighted sagittal MRI showing a cystic mass lesion at the level of S2 (black arrow). A : Case 1. B : Case 2.

cyst. Then, the CSF-like clear fluid was drained. The presence of several nerve roots was confirmed around the cyst and the cyst originated from ventral surface of the right S2 nerve root and compressed thecal sac. There was no free communication of CSF between the cyst and the intradural subarachnoid space, which was confirmed with an intraoperative Valsalva maneuver. Following a maximal level of cyst wall excision the cyst cavity and defect were covered using absorbable gelatin sponge (Gelfoam, Pharmacia & Upjohn, Kalamazoo, MI) and fibrin glue to prevent the postoperative CSF leakage that might occur. Surgical drainage was not inserted. The suture was done tightly in a layered fashion.

On histopathologic analysis, the irregular cystic wall composed of dense collagenous bundles including neural tissue along with vascular structures was observed. On immunohistochemistry stain (S-100 protein), the positive for neural tissue was observed (Fig. 3). On postoperative MRI scans, the previous cyst was totally removed. CSF leakage was not seen. The symptoms were improved immediately after the surgery (Fig. 4A).

Case 2

This 43-year-old woman, younger sister of the patient in Case 1, presented a 1-year history of perineal and perianal paresthesia which was also aggravated on positional change, such as standing, sitting and coughing, and relieved by lying down. One year prior to the outpatient visit, she was diagnosed with adenomyosis on gynecologic test for perineal pain. Then, she underwent total hysterectomy, but she was noted to persistently have perineal and perianal pain without any interval change. No past history of specific trauma or meningitis was noted. No abnormal findings were found on the preoperative hematologic studies. On neurological examinations, she complained of paresthesia in the right S2 and S3 dermatome. But, her motor weakness,

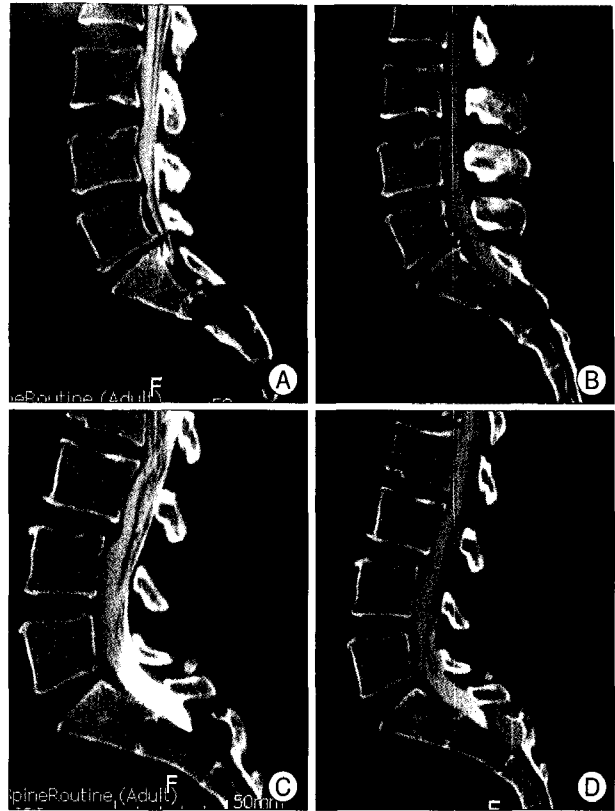


Fig. 2. Preoperative initial (A, C) and delayed myelo-computed tomography (CT) (B, D). A, B : Case 1. Preoperative myelo-CT showing no free communication between cyst and subarachnoid space (black arrow). C, D : Case 2. Preoperative myelo-CT showing delayed, but no free communication between cyst and subarachnoid space (black arrow).

bladder and bowel function were all intact. Electromyogram (EMG) showed no evidence of lumbosacral radiculopathy or peripheral neuropathy of lower extremities. On lumbosacral contrast enhanced MRI scans, there was a butterfly-shaped cystic lesion of $28 \times 13 \times 27$ mm in size, on the midline, particularly in the left neural foramen of S2, whose intensity was the same as CSF. No contrast-enhanced findings were observed (Fig. 1B). On initial myelo-CT scans, there was no free communication between cyst and subarachnoid space. On a 6-hour delayed myelo-CT scans, however, there was a low-density delayed filling rather than the contrast-enhancement of intradural subarachnoid space. In addition, there was the surrounding sacral bony erosion around this cyst (Fig. 2C, D).

Surgical treatment was done with same method. The presence of several nerve roots was confirmed around the cyst and the cyst originated from ventral surface of the right S2 nerve root and compressed thecal sac. But, unlike Case 1, there was a communication on the ventral wall of a cyst between the cyst and the intradural subarachnoid space. Because the cyst wall was too attenuated and too fragile, the closure of the communicating channel was impossible.

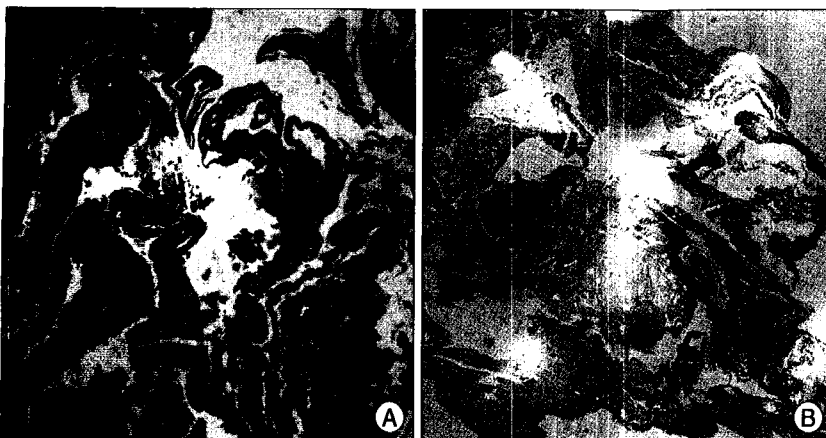


Fig. 3. Pathology. A : A photomicrograph showing irregular cystic wall composed of dense collagenous bundles including neural tissue along with vascular structures (H & E $\times 200$). B : A immunohistochemistry stain (S-100 protein) showing a positive for neural tissue.



Fig. 4. Postoperative magnetic resonance image (MRI). Postoperative T2-weighted sagittal MRI showing complete obliteration of cystic lesions. A : Case 1, B : Case 2.

Then the cyst cavity and defect were covered using Gelfoam and fibrin glue like Case 1.

Histopathologic findings of Case 2 were similar to those of Case 1. On postoperative MRI scans, the cyst was removed and CSF leaking was not seen. The symptoms were improved immediately after the surgery (Fig. 4B).

DISCUSSION

In 1938, Tarlov first described the cystic lesions in the sacral nerve root and coined the term "perineural cyst"¹⁵⁾. He found that these cysts were located in the perineurial space, between the endoneurium and perineurium at the junction of the posterior nerve root and its ganglion¹⁵⁾. Since then, in 1988, Nabors et al.¹⁰⁾ classified these cysts into three types : extradural meningeal cysts without spinal nerve root fibers (Type I); spinal extradural meningeal cysts with spinal nerve root fibers (Type II, Tarlov cyst); and spinal intradural meningeal cysts (Type III). In the current case, an operative inspection and histological examination revealed the presence of nerve root. The current case corre-

sponds to Type II in which the dilatation of posterior spinal nerve root sleeve is present.

To date, several hypotheses have been proposed to explain the etiologies of perineural cysts in the sacral region. Of them, the representative ones include inflammation within the nerve root cysts followed by inoculation of fluid, arachnoidal proliferation along and around the exiting sacral nerve root, breakage of venous drainage in the perineurium and epineurium secondary to hemosiderin deposition after trauma, and develop-

mental or congenital origin^{9,12,14,16)}. In cases which we have experienced, there was no past history of trauma, meningitis and surgery. During the fetal period, the dura mater was missing when it was developed. The subarachnoid membrane was escaped⁸⁾. Presumably, this might be responsible for the formation of the cyst. The current study may be the first report presenting symptomatic sacral Tarlov cyst with familial tendency.

Pathologically, sacral perineural cysts usually occur on the extradural components of sacrococcygeal nerve roots at the junction of dorsal root ganglion and posterior nerve roots and arise between the endoneurium and perineurium. And, the lining of these lesions contains nerve fibers and/or ganglion cells, which may surround the entire cavity¹⁶⁾. In the study of Voyazis et al.¹⁶⁾ nerve fibers were found in six (75%) of eight cases, and evidence of old hemorrhage in the form of hemosiderin was present in four (50%) of eight cases. This was attributed to the fact that Tarlov cysts are at different stages of evolution. In our cases, nerve fibers and vascular structures were present in both cases.

Surgical indications which several authors have reported until now include cases in which the size of a cyst is >1.5 cm and those in which the radiating pain is increased due to the postural changes and Valsalva maneuvers. In cases in which the positive filling defect sign indicating that the cyst does not actively fill with dye and compresses the other roots in myelography, the alleviation of clinical symptoms can be expected following the resection of a cyst^{1,13,16)}. These two cases that we are currently presenting herein met the criteria for operation indication mentioned above. Besides, we experienced the alleviation of symptoms following the surgery.

Sacral perineural cyst is not a common entity, whose exact etiology and pathophysiology remain unclear. No consensus has been reached on the ideal surgical treatment

modalities. Alternatively, the treatment options that have been reported until now include the method of diversing CSF flow (CT-guided percutaneous aspiration and lumbo-peritoneal shunt) and a direct microsurgical approach including the cyst wall excision combined with cyst wall imbrications together with defect repairing^{9,11,13,16}. Despite the absence of the active CSF communication between the cyst and the arachnoid space we covered the cyst cavity and defect using Gelfoam and fibrin glue to prevent the possible leakage of CSF. We observed no CSF leaking following the operations.

CONCLUSION

We report two operated cases of symptomatic sacral Tarlov cysts in one family. We have assumed that the genetic etiology is one of the important factors that are involved in the pathogenesis of Tarlov cyst.

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