

Ehlers-Danlos Syndrome Associated with Multiple Spinal Meningeal Cysts

—Case Report—

Mitsuo ISONO, Shigeaki HORI, Yohei KONISHI, Hidetomo KINJO,
Kenji KAKISAKO*, Ryuuichirou HIROSE*, and Takanori YOSHIDA*

*Department of Neurosurgery and *1st Department of Surgery, Oita Medical University, Oita*

Abstract

A 40-year-old female with Ehlers-Danlos syndrome was admitted because of a large pelvic mass. Radiological examination revealed multiple spinal meningeal cysts. The first operation through a laminectomy revealed that the cysts originated from dilated dural sleeves containing nerve roots. Packing of dilated sleeves was inadequate. Finally the cysts were oversewed through a laparotomy. The cysts were reduced, but the postoperative course was complicated by poor wound healing and diffuse muscle atrophy. Ehlers-Danlos syndrome associated with spinal cysts may be best treated by endoscopic surgery.

Key words: spinal meningeal cyst, Ehlers-Danlos syndrome

Introduction

Ehlers-Danlos syndrome (EDS) is a rare hereditary disorder of collagen metabolism, of which nine types and several subtypes have been recognized.²⁾ Patients with EDS type IV, the so-called "vascular type," sometimes show neurosurgical manifestations such as subarachnoidal hemorrhage or carotid cavernous fistulae.¹⁾ Patients with other types may suffer from neurological symptoms due to spinal deformities.⁵⁾ Other neurosurgical manifestations are rare. We describe a unique case of EDS associated with multiple meningeal cysts.

Case Report

A 40-year-old female was admitted on June 10, 1996, because of a pelvic mass. A dermatological diagnosis of EDS type VI was made when she was 14 years old. She had suffered from her right inguinal herniation since 20 years old, but had not undergone the surgery. She also suffered from intermittent diarrhea for several years previously. Several months before admission, she suffered from left lower abdominal pain. Radiological examination revealed a

cystic mass arising from the sacral spinal canal. Thereafter, her abdominal pain gradually increased and was radiating to her left leg at admission.

On admission, her height was 168 cm and her body weight was 56 kg. Physical examination revealed mild scoliosis, joint hypermobility, hyperextensible skin, and arachnodactyilia. There was a large mass in the right inguinal region suggesting inguinal herniation and slight tenderness in her left lower quadrant abdomen which radiated to her left leg. Neurological examination revealed slight sensory disturbance in the left L4-S1 area.

Radiography showed no apparent bone destruction of the sacrum. Computed tomography (CT) revealed an enlarged sacral spinal canal and a cystic mass extending from the anterior foramina down into the pelvis bilaterally (Fig. 1). Magnetic resonance (MR) imaging disclosed that the cyst contents had the same intensity as cerebrospinal fluid. Myelography through a lumbar puncture did not reveal direct communication between the cysts and the subarachnoid space, but CT performed 3 hours later showed influx of the contrast medium into both cysts (Fig. 2). MR imaging of the cervical and thoracic spine showed multiple dural ectasia in various regions (Fig. 3). We considered that her symptoms including the low abdominal pain radiating to the left foot might be due to compression of

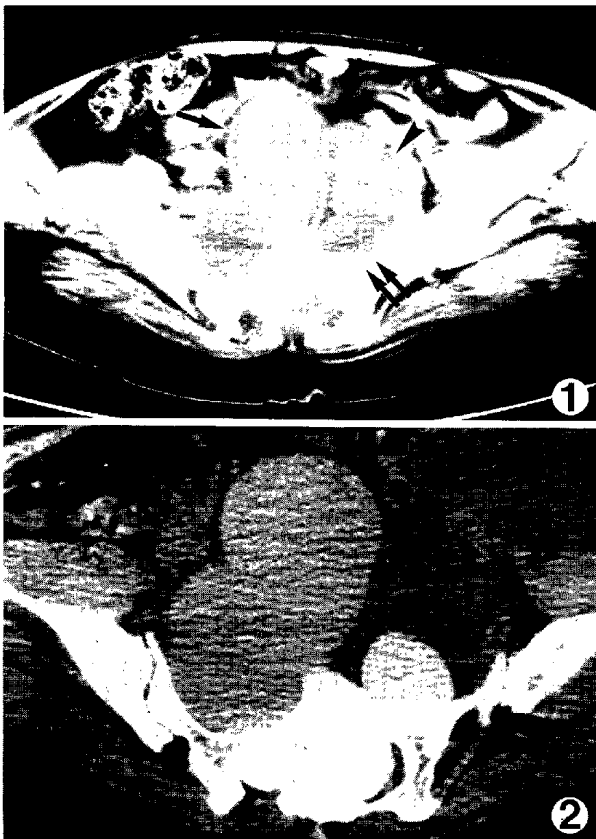


Fig. 1 Computed tomography scan showing the dilated sacral spinal canal and a large cyst in the right (arrow) and a small cyst in the left (double arrows) pelvis, as well as dilated bladder (arrowhead).

Fig. 2 Computed tomography scan performed 3 hours after direct myelography showing influx of contrast medium into both cysts.

the spinal nerve roots by the cysts, although the cyst on the left side was small.

Laminectomy was selected for the first operation on July 30, because of the potential risk to damage abdominal organs or vessels and problems with wound healing through laparotomy. There was small amount of quite loose subcutaneous tissue. The bulging sacral laminae were very thin and a L4-S5 laminectomy disclosed extremely thin dura mater without epidural connective tissue. Observation through the enlarged spinal canal revealed an enlarged dural sleeve at the right L-5 with a diameter of 3 cm and an enlarged dural sleeve of the left S-1 with a diameter of 2 cm (Fig. 4). Intraoperative radiographs confirmed the slow influx of contrast medium into the cysts through these sleeves. However, we could not directly identify the ostium of the cysts. Both enlarged sleeves were packed with

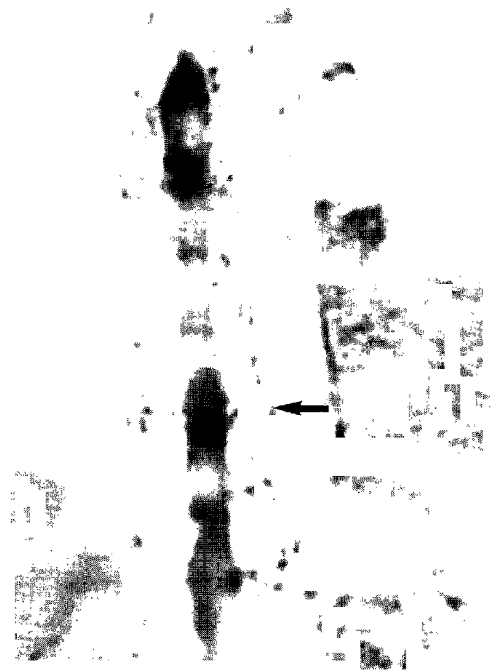


Fig. 3 Coronal T₂-weighted magnetic resonance image showing the dilated dural sac (arrow).

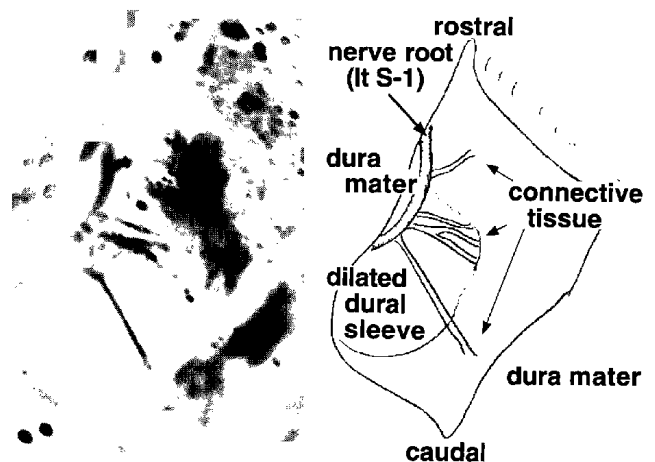


Fig. 4 Intraoperative microscopic view (left) showing the dilated dural sleeve and left S-1 nerve root. Schema of the microscopic view (right).

fatty tissue.

Postoperatively, her low abdominal pain radiating to the left leg did not diminish. MR imaging 6 weeks after the operation showed the size of the cysts had not changed and CT taken after intrathecal injection of contrast medium revealed persistent communication between the cysts and the subarachnoid

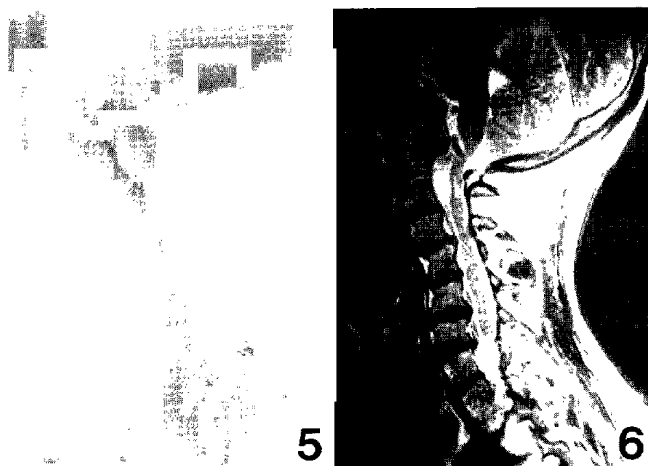


Fig. 5 Tomogram of cervical spine showing spondylolisthesis at C2-3 intervertebral level and the hypoplastic atlas.

Fig. 6 T₂-weighted magnetic resonance image showing spondylotic changes of the cervical spine, severe compression of the cord at C3-4 intervertebral level, and the high intensity area in the upper cervical cord.

space. Laparotomy was performed on September 24, and the remaining cysts were opened. The nerve root was running within the dorsal wall of both cysts and Valsalva maneuvers revealed influx of cerebrospinal fluid into both cysts. The cyst walls were oversewn with packing connective tissue inside. The right inguinal hernia was treated during the same operation.

These procedures resulted in reduction of both cysts. However, her postoperative course was rather complicated. She suffered from new pain in the bilateral inguinal and femoral areas, possibly secondary to the suture procedure of the cysts and inguinal hernia. Wound healing was quite bad and the skin wound took 5 months to heal. Dehiscence of the muscle and subcutaneous layer persisted as an abdominal wall herniation.

She gradually developed motor and sensory disturbances in both extremities and finally became tetraparetic with bladder and anorectal dysfunction. Detailed examinations indicated that these neurological disturbances were attributable to severe cervical spondylolisthesis associated with hypoplasia of the atlas (Figs. 5 and 6). Disuse muscle atrophy resulting from long-term bed-confinement due to pain and delayed wound healing may have accelerated these conditions. She gradually improved during physical therapy with a neck brace. Eight months after the second operation, she was discharged walking with the assistance of a cane and

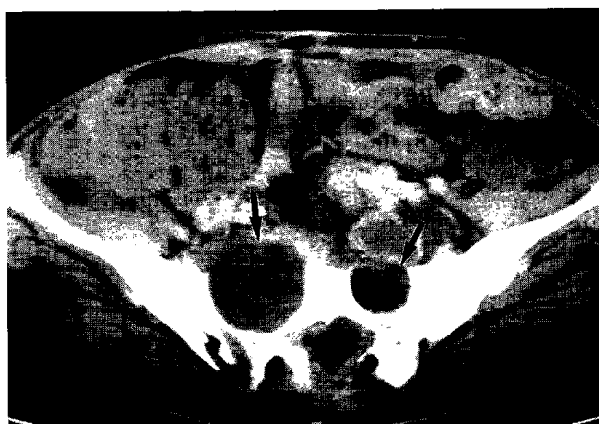


Fig. 7 Computed tomography scan performed 6 months after the second surgery showing reduced size of the cysts (arrows).

returned home. Follow-up examinations showed no regrowth of the cysts (Fig. 7).

Discussion

The classification of spinal cystic lesions is rather confusing with a variety of terms used. Extradural cysts of the sacral region have been described as "perineurial cysts,"^{7,13)} "meningeal diverticula,"¹⁾ "occult intrasacral meningocele,"⁴⁾ and others.

Spinal meningeal cysts have been classified according to clinical and histological findings into three groups: extradural meningeal cysts without spinal nerve root fibers (type I); extradural meningeal cysts with spinal nerve root fibers (type II); and intradural meningeal cysts (type III).⁶⁾ According to these criteria, "sacral meningocele" is type I and "perineurial cysts" and "spinal nerve root diverticula" are type II.⁶⁾ The clinical and intraoperative findings suggest our case might belong to type II, although histological evidence is not available.

Hereditary disorders of connective tissue include association of spinal meningeal cysts, mainly type I meningeal cysts, with Marfan's syndrome.^{10,12)} Patients with Marfan's syndrome have a high incidence of dural ectasia.⁹⁾ Dural ectasia is thought to be the precursor to the development of meningeal cysts and is thought to result from pulsatile pressure of cerebrospinal fluid upon a pathologically elastic dural membrane in patients with Marfan's syndrome.

Since such conditions as pathological elasticity in the connective tissue, including the dural membrane, might be similar in patients with EDS, an association with meningeal cysts is certainly possible. Our case showed multiple dural ectasia in vari-

ous regions, which might be considered as the precursors of meningeal cysts. However, the association of meningeal cysts in patients with EDS has not been found. This rarity might be partly due to the lower incidence of EDS, estimated at 1:200,000 living birth,⁸⁾ compared to the incidence Marfan's syndrome, 4 to 6:100,000 living birth.³⁾ Moreover, there might be possibility of overlooking, because meningeal cysts usually show only common symptoms such as pain or constipation.

Different strategies can be used for the treatment of cysts.⁵⁾ Type I meningeal cysts have a pedicle, so can be treated by closing the ostium between the cyst and the subarachnoid space. Type II meningeal cysts seldom have a pedicle, so excision of cyst with oversewing of the wall or complete resection are required.⁶⁾ The problems in the surgical treatment of patients with EDS originate in the fragility of tissue, including abdominal organs and major vessels. Therefore, laparotomy in our patient had greater risks for wound healing and damage to abdominal organs or vessels. We first tried to pack the dilated dural sleeves through laminectomy, resulting in being insufficient. The reason might be that we could not identify the entire enlarged dural sleeves and the ostium of the cysts directly. The inadequate result strongly supports the recommended strategy. The subsequent laparotomy was successful, but lead to problems with wound healing, complicated by period of bed confinement. Since patients with EDS have loose and pathologically elastic connective tissue, including ligamenta and paravertebral supportive tissue, disuse muscle atrophy made the instability of cervical spine worsen in our patient, resulting in neurological deterioration. Although the hypoplasia of atlas observed in our patient might be uncommon, disorders of spine, such as spondylosis or spondylolisthesis, are common, besides scoliosis, in patients with EDS.³⁾ Therefore, in patients with EDS associated with spinal cysts, close examinations of general condition should be required before the operation and less invasive surgery such as endoscopic surgery should be considered.

References

- 1) Basauri L, Hudson H, Bradales A: Diverticuli of the nerve root sheaths. Case report. *J Neurosurg* 31: 680-682, 1969
- 2) Beighton P, Paep A, Danks D, Findori G, Gedde-Dahl T, Goodman R, Hall JG, Hollister DW, Horton W, Mckusick VA, Opitz JM, Pope FM, Pyeritz RE, Rimo DL, Sillence D, Spranger JW, Thompson E, Tsipouras P, Vrijioen D, Winship I, Young I: International nosology of heritable disorders of connective tissue, Berlin, 1986. *Am J Med Genet* 29: 581-594, 1988
- 3) Goldman AB: Collagen disease, epiphyseal dysplasia, and related conditions, in Resnick D, Niwayama G (eds): *Diagnosis and Treatment of Bone and Joint Disorders*. Philadelphia, WB Saunders, 1988, pp 3374-3441
- 4) Lamas E, Lobato RD, Amor T: Occult intrasacral meningocele. *Surg Neurol* 8: 181-184, 1977
- 5) McMaster MJ: Spinal deformity in Ehlers-Danlos syndrome. Five patients treated by spinal fusion. *J Bone Joint Surg Br* 76: 773-777, 1994
- 6) Nabors MW, Pait TG, Byrd EB, Karim NO, Davis DO, Kobrine AI, Rozzoli HV: Updated assessment and current classification of spinal meningeal cysts. *J Neurosurg* 68: 366-377, 1988
- 7) North RB, Kidd DH, Wang H: Occult, bilateral anterior and intrasacral meningeal and perineurial cysts: case report and review of the literature. *Neurosurgery* 27: 981-986, 1990
- 8) Pinnell SR: Ehlers-Danlos syndrome, in Berson PB (ed): *Cecil Textbook of Medicine*. Philadelphia, WB Saunders, 1979, pp 2054-2055
- 9) Pyeritz RE, Fishman EK, Bernhardt BA, Siegelman SS: Dural ectasia is a common feature of the Marfan syndrome. *Am J Hum Genet* 43: 726-732, 1988
- 10) Raftopoulos C, Poerard GE, Retif C, Braude P, Brotchi J: Endoscopic cure of a giant meningocele associated with Marfan's syndrome. *Neurosurgery* 30: 765-768, 1992
- 11) Schievink WI, Limburg M, Oorthuys JWE, Fleury P, Pope FM: Cerebrovascular disease in Ehlers-Danlos syndrome Type IV. *Stroke* 21: 626-632, 1990
- 12) Schneider MB, Dittmar S, Boxer RA: Anterior sacral meningocele presenting as a pelvic/abdominal mass in a patient with Marfan syndrome. *J Adolesc Health* 14: 325-328, 1993
- 13) Tarkov IM: Spinal perineurial and meningeal cysts. *J Neurol Neurosurg Psychiatry* 33: 833-843, 1970

Address reprint requests to: M. Isono, M.D., Department of Neurosurgery, Oita Medical University, 1-1 Idaigaoka, Hasama-machi, Oita-gun, Oita 879-5593, Japan.