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Terminal Myelocystocele
— A Case Report —

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Terminal myelocystocele is a rare form of occult spinal dysraphism in which the hydromyelic caudal spinal cord and the subarachnoid space are herniated through a posterior spina bifida. A 1.5 month old boy presented with a large lumbosacral mass and urinary incontinence. The magnetic resonance imaging, operative findings and pathological findings revealed a low lying conus with a dilated central canal dorsally attached to the subcutaneous tissue. Ventral subarachnoid space was enlarged and herniated through the laminar defect of the sacrum. The lesion was typical of a terminal myelocystocele. The clinical features are different from those of myelomeningocele in many aspects. Though the incidence is low, terminal myelocystocele should be included in the differential diagnosis of congenital lesions presenting as a lumbosacral mass.

Key Words: *Terminal myelocystocele, Spinal dysraphism, Low lying conus, Dilated central canal, Lumbosacral mass*

INTRODUCTION

Myelocystocele is defined as an occult spinal dysraphism with a localized, cystic dilatation of the central canal of the spinal cord which is herniated through a posterior spina bifida. Terminal myelocystocele is truly an anomaly of the caudal cell mass and can be associated with anomalies of the anorectal system, lower genitourinary system, and vertebrae, such as anal atresia, cloacal exstrophy, lordosis, scoliosis, and partial sacral agenesis (McLone and Naidich, 1985; Warf et al., 1993; McLone and Dias, 1994). Cervicothoracic myelocystoceles are of unknown cause, but clearly not the result of disturbances of the caudal cell mass, so that these lesions should not be confused with terminal myelocystocele (Suneson and Kalimo, 1979). There have been a few articles written in English which illustrate the radiologic or operative findings of terminal myelocystocele (Lemire et al., 1971; Hayden et al., 1973; Schmitt and Kawakami, 1982; McLone and Naidich, 1985; Peacock and Murovic, 1989; McLone and Dias, 1994). To our knowledge, this is the first reported case of a terminal myelocystocele in Korea.

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CASE REPORT

This baby boy was a 3.6 kg product of 39-week pregnancy and of a normal spontaneous vaginal delivery. At birth, a 7 × 4.5 cm lumbosacral skin-co-

vered mass occupied and obliterated the intergluteal cleft. The anterior abdominal wall and genitalia were normal.

Admission

The patient was brought for neurosurgical repair of the lumbosacral mass at 1.5 months of age. On physical examination, the soft mass was 9 × 7 × 2.5 cm in size with normal skin on the surface. The head circumference was 38 cm, 50th percentile. There was a patulous anus. Neurologically, motor function was grossly intact without any foot deformities. Saddle anesthesia and urinary incontinence were noted. Plain spinal radiographs showed normal cervical, thoracic, and lumbar vertebrae ; whole sacral posterior spina bifida and wide interpedicular distance of the sacrum. Radiographs of pelvis demonstrated normal symmetric pelvis and hip joints. Cranial sonography and computed tomography (CT) showed normal ventricles and brain parenchyma without Chiari malformation. Spinal ultrasonography showed a huge lumbosacral cystic lesion with double compartments which were connected with the spinal canal. A low-lying herniated cord was attached to the cystic mass. The subarachnoid space, located ventral to the dorsal cyst which was the continuation of the central canal, was bulged out of the spinal canal. Voiding cystourethrography showed vesicoureteral reflux in the left side and the dilated bladder. A T1-weighted magnetic resonance (MR) image (Fig. 1) revealed a terminal myelocystocele which contained a trumpet-like flaring of the distal spinal cord central canal and a meningocele or dilated subarachnoid space located around the dilated central canal, which bulged into the subcutaneous region. The signal intensity of fluid in the dilated central canal was slightly higher than that of subarachnoid space in T1-weighted image. The high signal intensity in the area of soft-tissue enlargement dorsal and superior to the cystic lesion was compatible with the subcutaneous fat tissue. Urodynamic study revealed an areflexic bladder with low compliance and vesicoureteral reflux of grade III/V in the left side.

Operation

A neurosurgical procedure was performed in February, 1991, at 1.5 months of age. At the caudal part of the sac, the terminal cyst, which was actually the dilated terminal central canal, was opened and slightly xanthochromic cerebrospinal fluid (CSF) gushed out. The myelocystocele itself had septa and had no communication with the subarachnoid space. The spinal cord was dilated and continued to the dorsal cyst which is the dilated central canal (Fig. 2B). After aspiration of clear CSF from the meningocele, the dura and arachnoid above the dilated central canal were opened. The roots traversed the sac from the cord in cephalad direction. On the stimulation of the distal roots, ankle plantar flexion responses were evoked. At the caudal end of the meningocele, the arachnoid was reflected from the wall of the meningocele to the pia-arachnoid of the spinal cord. The cord itself bulged at the meningocele, where it was

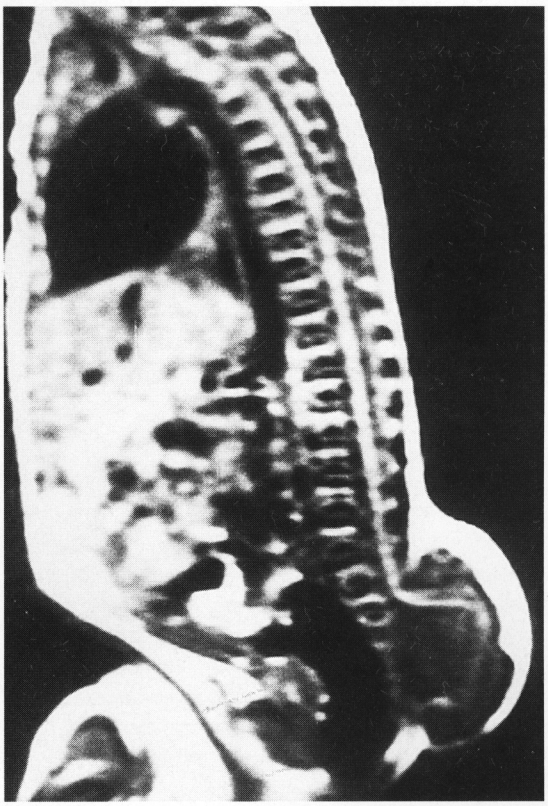

Fig. 1. T1-weighted magnetic resonance image, sagittal view, shows an expanded, syringohydromyelic spinal cord that appears to terminate at the mid-sacral level. At the cord terminus, the central canal opens into a large cyst. At surgery, this terminal cyst was found to communicate with the dilated central canal of the spinal cord, resulting in the classification of this anomaly as a myelocystocele. The meningocele is also visualized ventral to the dorsal cyst which is the continuation of the central canal.

attached to the extraarachnoid subcutaneous fat tissue. The spinal cord was untethered at the junction with the fat tissue. The pial reconstruction was performed along the cut surface (Fig. 2C). An extradural

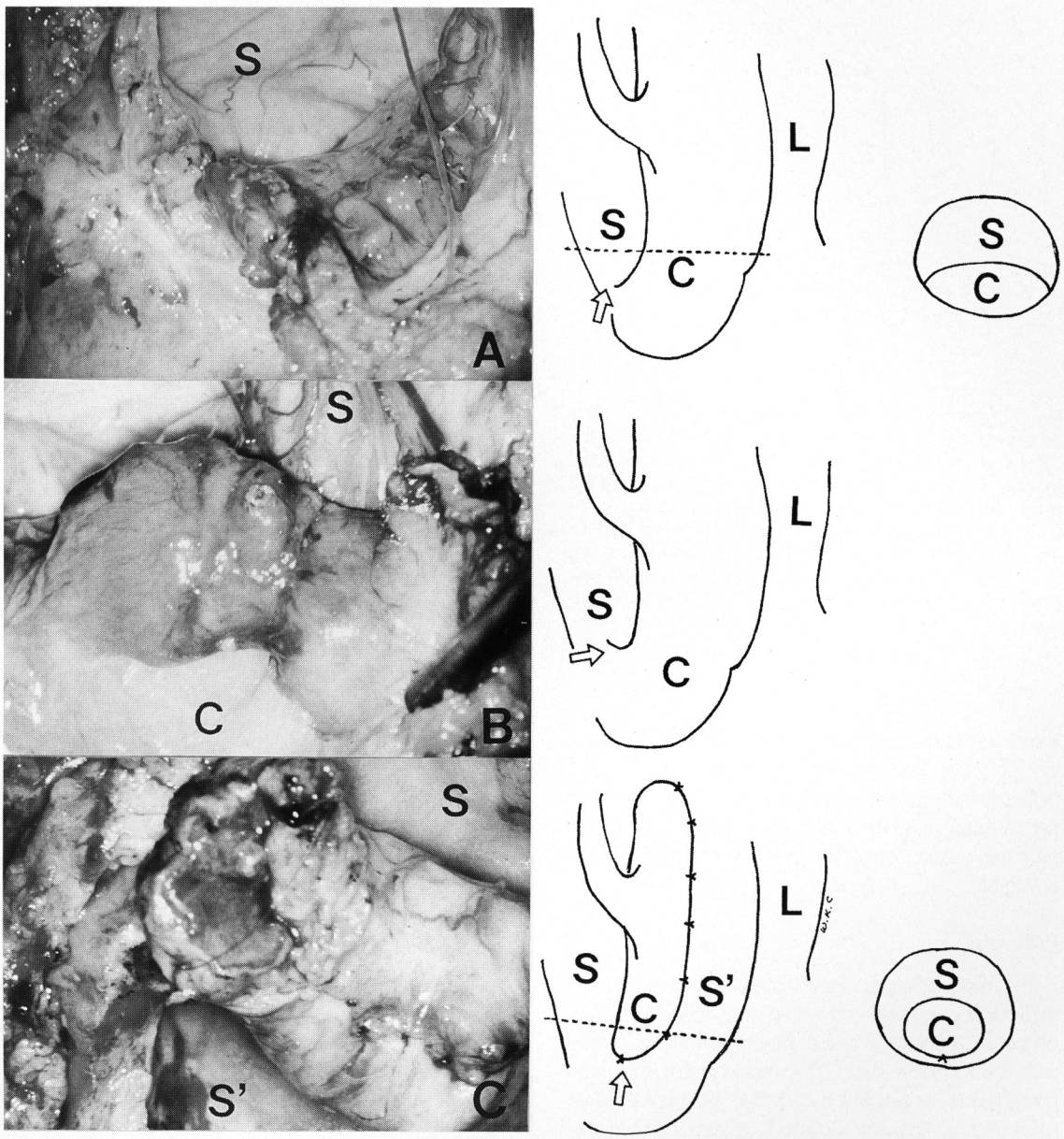

Fig. 2. Operative photographs and schematic illustrations. A : The subarachnoid space was entered from the infero-dorsal aspect. Pial vessels and rootlets are seen. B : After circumferential incision of the wall of dorsal cyst (dilated central canal, untethering), the edge of the spinal cord is shown. There was no communication between the dorsal cyst and the ventral subarachnoid space. The suction tip is on the surface of the dilated central canal. C : For the pial reconstruction, the free edges of the thinned out spinal cord were approximated. The most caudal part was being recontructed. S : subarachnoid space, C : central canal, L : lipoid tissue, S' : subarachnoid space posterior to the reconstructed spinal cord. For A and C, schematic cross sectional drawings at the level of dashed lines are shown. Arrows indicate the directions of view for each photograph.

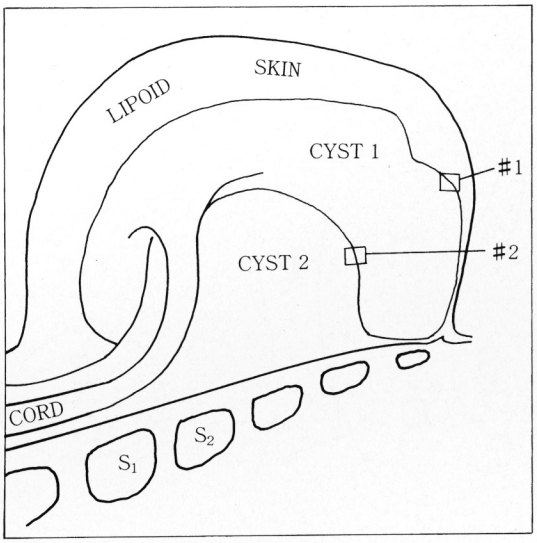

Fig. 3. Schematic drawing of the operative findings. The content of cyst 1 was clear xanthochromic fluid and the content of cyst 2 was watery clear fluid. The pathologic findings of specimen taken at the site #1 was 'neuroglial tissue with ependymal lining, meningeal and adipose tissues'. The pathologic findings of specimen taken at the site #2 was 'fibroadipose tissue with peripheral nerve twigs'. The cyst 1 was the dilated central canal (cystocele) and the cyst 2 was the extension of spinal subarachnoid space (meningocele).

fibrous band crossing the midline at the upper part of the defective laminae was resected and the next higher intact lamina was removed to expose the normal dura mater and its posteroinferior continuation over the meningocele. The dura was closed without any graft.

Pathology

The histopathologic examination revealed that the myelocystocele wall consisted of ependyma-lined dysplastic neuroglial tissue mixed with fibroadipose and meningeal tissues. The meningocele wall was fibroadipose tissue which included peripheral nerve twigs (Fig 3). Analysis of cystic fluid from the myelocystocele revealed 10 red blood cells/mm^3, 17 white blood cells/mm^3. The values of protein and glucose were 576 mg/100 ml and 97 mg/100 ml, respectively. Analysis of CSF from the meningocele revealed no cell and the values of protein and glucose were 58 mg/100 ml and 57 mg/100 ml, respectively.

Postoperative course

The patient tolerated the procedure well. There was no definite difference between the two postoperative MR images which had been taken at ages 2 and 3 (Fig. 4). They showed a low-lying conus down to S2 area. There was no syrinx or terminal myelocystocele. Meningocele also disappeared. The bladder was enlarged. At 3 years after surgery, he could walk and run well without any weakness or skeletal deformities, without complaining of any pain. Though his bowel control was intact, he had an intermittent unrinary incontinence.

DISCUSSION

Terminal myelocystocele consititutes 4-8 % of lumbosacral occult spinal dysraphism (Lemire et al., 1971

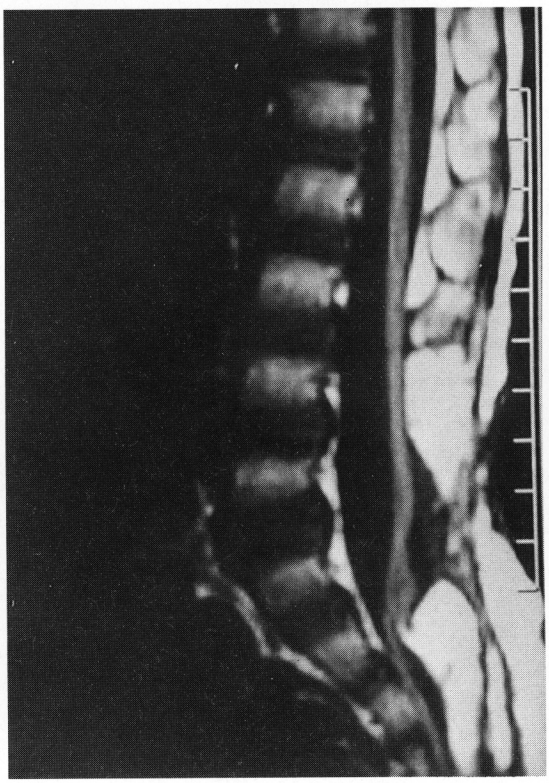

Fig. 4. A T1-weighted magnetic resonance image taken at 3 years after surgery shows a low lying conus. The previous meningocele sac and the dorsal cyst disappeared. Though the spinal cord was tethered radiologically, it will be untethered if the clinical findings suggest 'symptomatic retethering'.

; McLone and Naidich, 1985). Epidemiologically, myelocystoceles arise sporadically ; there is no known familial incidence (Carey et al., 1978 ; McLone and Naidich, 1985).

This lesion consists of a skin-covered lumbosacral spina bifida, an arachnoid-lined meningocele directly continuous with the spinal subarachnoid space, and a low-lying hydromyelic spinal cord that traverses the meningocele and forms a distal sac that does not communicate with the subarachnoid space. The terminal cyst is lined by ependyma and dysplastic glia, and is directly continuous with the dilated central canal of the cord, probably representing a ballooned terminal ventricle (McLone and Naidich, 1985 ; McLone and Dias, 1994).

Lemire et al. (1971) found only two cases of terminal myelocystocele in a review of 31 skin-covered lumbosacral masses. In another series of 48 skin-covered lumbosacral masses, McLone and Naidich (1985) found only two terminal myelocystoceles. Hayden et al. (1973) presented two cases of terminal myelocystocele associated with exstrophy of the cloaca, and Schmitt and Kawakami (1982) reported another example. In a case of bladder exstrophy presented by Russell (1939), the lesion was not called terminal myelocystocele. Recently Peacock and Murovic (1989) added two cases with detailed MR appearance. This is an entity which has probably been overlooked in the past due to inadequate diagnostic technique. This entity should be considered when pediatric lumbosacral masses are evaluated. There are undoubtedly additional cases in the literature incorrectly described as other disease entities.

Terminal myelocystoceles are thought to arise during the period of secondary neurulation from the caudal mass (McLone and Naidich, 1985 ; Peacock and Murovic, 1989 ; McLone and Dias, 1994). McLone and Naidich (1985) postulated that the terminal myelocystocele arises because CSF is unable to exit from the central canal. This dilates the terminal ventricle, disrupting the dorsal mesenchyme but not the surface ectoderm. Thus, a spina bifida develops beneath the intact skin. Continued growth of the terminal cyst by accumulation of CSF distends the surrounding arachnoid lining of the distal thecal sac, causing formation of a meningocele. Progressive distension of the distal cord causes it to bulge caudally below the end of the meningocele into the extraarachnoid space, where it is covered by fat. The bulk of cyst also bulges cephalad to expand the distal cord producing trumpet-like flaring and prevents ascent of the cord, producing tethered cord. Scattered data also suggest a possible relationship to teratogens such as hydantoin, ioperamide hydrochloride, and retinoic acid, although the exact etiology of this entity is not known (Shenefelt, 1972 ; Carey et al., 1978 ; Schmitt and Kawakami, 1982).

The nature of the underlying CSF disturbance is uncertain. It is important to note that hydrocephalus is extremely uncommon in patients with terminal myelocystoceles, suggesting that the disturbance of CSF dynamics is local rather than global and the patients have normal intellectual potential (McLone and Dias, 1994).

Clinically the lumbosacral cystic, skin-covered mass is nearly always visible at birth and varies in size from small to huge (Hayden et al., 1973 ; McLone and Naidich, 1985). The mass typically occupies and obliterates the intergluteal cleft and extends upward from the perineum for a variable distance. The skin overlying the mass may appear normal or may exhibit hemangioma, nevus, or hypertrichosis (Schmitt and Kawakami, 1982 ; McLone and Naidich, 1985). A few patients seem to have deficits at birth. Others are born with normal lower extremities, but lose function over time, probably as a result of the tethered spinal cord (McLone and Naidich, 1985).

Until the advent of MR imaging, metrizamide myelography followed by CT scanning in an axial plane was used to evaluate lumbosacral masses. Now, MR imaging is the radiographic modality of choice for patients with myelocystocele, and also delineates a number of associated abnormalities. Imaging studies demonstrate direct continuity of the meningocele with the subarachnoid space and the presence of a cyst which is continuous with the central canal of the spinal cord. Because the cyst does not communicate with the meningocele, myelography, if performed, will demonstrate only the meningocele. Imaging studies show that the clinically apparent mass is a second cyst that is thin-walled and has no internal structure. The ependyma-lined cyst is frequently the larger of the two cysts ; it is typically situated posteriorly and inferiorly to the meningocele and occasionally extends rostrally outside the meningocele (Peacock and Murovic, 1989).

McLone and Naidich (1985) suggested that tethered cord and disruption of the caudal motor segments produce symptoms that may be present at

birth or may appear later to progress. They experienced two patients with terminal myelocystocele. One of them showed improvement of lost neurological function after operation. Based on their experience, they presented terminal myelocystocele as a distinct, surgically treatable malformation with the need for early diagnosis and intervention. In our case the patient presented with a lumbosacral mass at birth and urinary incontinence without low extremity deformities or skeletal deformities. After operation the patient showed good motor development. Postoperative MR images revealed that the lesions had been well repaired. Low lying conus and enlarged bladder, however, were shown on MRI. He has walked and run well till 3 years after operation. He will be observed periodically as to whether any symptoms occur from the radiological tethered cord.

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