

Occult Intrasacral Meningocele Associated With Tethered Cord Syndrome

HAYATI ATABAY, YUSUF ERŞAHİN SAFFET MUTLUER, HASAN MİRZAI

Division of Paediatric Neurosurgery, Ege University Medical School, Bornova-İzmir, Turkey

Abstract : True occult intrasacral meningocele is a rare abnormality. A case associated with spinal cord tethering which was identified with magnetic resonance imaging (MRI) in an adolescent is presented. MRI's role in the diagnosis is discussed.

Key Words : Intrasacral cyst, occult meningocele, magnetic resonance imaging

INTRODUCTION

Occult intrasacral meningocele refers to a dilated intrasacral extension of the thecal sac that does not contain neural elements (1,12). It is composed of fibrous tissue resembling dura mater and attached to the caudal termination of the dural sac distal to the S2 vertebral body, usually by a narrow pedicle. Free cerebrospinal fluid (CSF) flow from the subarachnoid space to the meningocele is usually present (1,10,15). Enderle (3) first demonstrated occult sacral meningocele with myelography and introduced this term in 1932. The first surgically verified case was reported by Walker (14) in 1944, and Rengachary et al. (11) reported in 1981 that there were fewer than 50 cases found in the literature.

We report a unique case of tethered cord, associated with occult intrasacral meningocele which had been followed-up as familial spastic paraplegia since 1984.

CASE REPORT

A 12-year-old boy was admitted to the Division of Paediatric Neurosurgery with bowel and urinary incontinence, bilateral progressive leg weakness and

foot deformities which had been present since infancy. For 8 years he had been followed-up with the diagnosis of familial spastic paraplegia by the Department of Neurology where, because of the progressive neurological deterioration and foot deformities, spinal MRI was done to investigate the etiology. After determination of spinal cord tethering, he was referred to our department. General physical and neurological examination disclosed minimal tenderness over the sacrum on palpation, bilateral ankle deformity and pes echino varus, spastic paraparesis, bilateral absence of ankle jerks, hypoesthesia below L3 and decreased sphincter tone. MRI disclosed low-lying conus medullaris and thickened filum terminale and also some signal intensity as CSF, arising from the most caudal aspect of the dural sac below the S2 level (Fig. 1a,b). Plain X-ray films revealed sacral spina bifida and widened sacral canal (Fig. 2). At operation L5 laminectomy was performed and an intact dural sac was detected. Sacral dissection exposed a large cyst filling the enlarged sacral spinal canal. The cyst wall was thin and bluish. A narrow fistulous communication between the lumbosacral meningeal sac and the cyst was detected. The dura was incised and a tethered filum terminale identified and then sectioned.

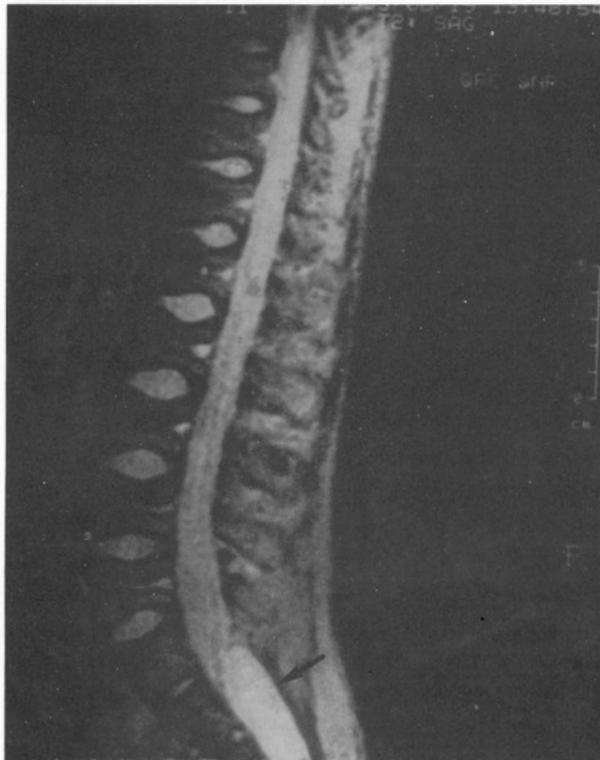


Fig. 1 : (a) T1-weighted and (b) T2-weighted MRIs in the sagittal plane demonstrating low lying conus medullaris and thickened filum terminale and intrasacral meningocele (arrow). The cyst has the same high signal intensity as the cerebrospinal fluid.

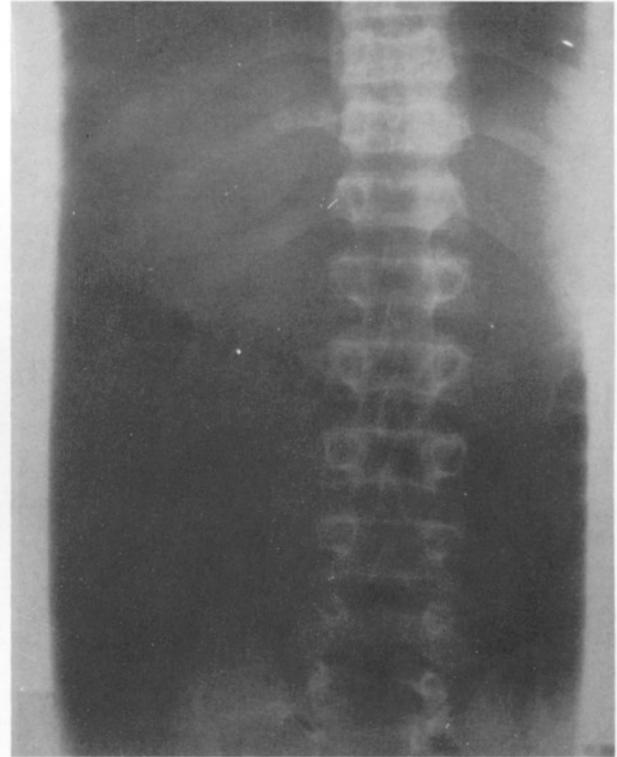


Fig. 2 : A-P plain radiograph showing expansion of the sacrum and the bifid first sacral spinous process.

As the size of the cyst remained the same, the dura was closed with a suture, and the cyst was incised. No nerve roots were present within the cyst cavity or cyst wall. The fistulous tract between the lumbosacral subarachnoid space and the cyst was ligated and the walls of the cyst were removed (Fig. 3). The postoperative course was uneventful. The patient was free of back pain one month after surgery, however neurological signs had not changed.

DISCUSSION

Occult intrasacral meningocele is a rare abnormality (4,7,8,9). Congenital, ischaemic degenerative, traumatic and iatrogenic causes have been suggested for meningeal and perineural cyst occurrence (1,7,12). As in ours, in some cases, the presence of other associated congenital anomalies supports the congenital nature of occult intrasacral meningocele (4,9). Nabors et al. (10) classified spinal meningeal cysts in three categories:



Fig. 3 : Postoperative T1-weighted MRI in the sagittal plane demonstrating that the intrasacral cyst has disappeared and the conus medullaris is elevated.

cult intrasacral meningocele is difficulty in visualization of the sacrum or not examining the sacrum carefully during studies (4). In our case, visualization of the sacrum on the plain X-ray was not complete, the MRI evaluation was insufficient, and axial plane images were not taken in the sacral area.

Clinical presentation of tethered filum terminale and sacral meningocele may include gait disturbance, foot deformity, bowel and bladder complaints, back or leg pain due to tethering and/or compression of the conus medullaris and lumbosacral roots (1,2,5,6,9,10,13,15). A sacral meningocele, probably due to hydrodynamic effects of the CSF in it, enlarges slowly and usually does not become symptomatic until adult life (2,4,15). A valvular type of communication between the thecal sac and the cyst would explain the high pressure of the cyst which causes compression of the sacral roots (6,9).

In our case, we think that the absence of lumbar skin abnormality, sacral localization of occult spinal dysraphism and insufficient familial history caused the patient to be followed-up as familial spastic paraplegia. However, as MRI disclosed the low-lying conus medullaris, thickened filum terminale and accompanying occult intrasacral meningocele, we think that both these pathological conditions were probably responsible for the clinical progression.

Some authors do not suggest surgery for sacral meningocele in asymptomatic cases. However, some of these cysts may grow in size by a hydrodynamic mechanism and become symptomatic (1,6,15). In our opinion, an asymptomatic cyst can be excised during surgery for another pathology in an a nearby area. The sacral and coccygeal nerve roots should be separated from the entire cyst. Surgical treatment consists of ligating and dividing the pedicle and excising the meningocele sac (4,15). Occult intrasacral meningocele is a rare abnormality. In some cases, it may cause low back and leg pain and can imitate lumbar disc herniation (1,4,6). Various neurological symptoms and signs, and also foot deformities can be seen. The sacral area should be examined carefully and if feasible, MRI should be used for further investigation in cases with unexplainable neurological deficits, orthopaedic deformities in the lower limbs and persistent low back and leg pain when conventional radiological studies do not reveal the etiology.

- Type I : Extradural meningoceles without spinal nerve root fibers
 - A Extradural meningeal cyst (Extradural arachnoid cyst)
 - B Sacral meningocele (Occult sacral meningocele)
- Type II : Extradural meningoceles with spinal nerve root fibers (Tarlov's perineural cyst)
- Type III: Spinal intradural meningoceles (Intradural arachnoid cyst)

Surgical confirmation of the absence of neural elements is important to distinguish between intrasacral meningocele and perineural cyst (7,12). Plain X-rays of the sacrum usually show enlargement of the sacral spinal canal with thinning of the overlying lamina and scalloping of the sacral vertebral bodies (4,6,8,10,14). Visualization of the cord and thecal abnormalities are limited by myelography and computerized tomography (2,6). MRI appears to be the single most useful and definitive study to identify an intraspinal cystic mass and other associated abnormalities (2,6,10). One of the most frequent problems in the diagnosis of oc-

Correspondence : Hayati Atabay, M.D.
Ali Çetinkaya Bulvarı
Doktorlar Sitesi
Alsancak İzmir, Turkey
Phone: (232) 463 8524

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