Anterior sacral meningocele with an unusual presentation

Case report

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A case of familial anterior sacral meningocele associated with a dermoid tumor is reported. This patient presented with recurrent aseptic meningitis. The role of computerized tomography following metrizamide myelography in the diagnosis of this lesion is discussed.

KEY WORDS  •  aseptic meningitis  •  dermoid cyst  •  familial anomaly  •  anterior meningocele  •  myelography  •  computerized tomography

Anterior sacral meningoceles (ASM’s) are rare examples of spinal dysraphism. They are created by herniation of the thecal sac through a bone defect in the anterior sacral wall. A recent review of this entity16 plus five additional case studies2,12,13 have placed the total number of reported occurrences at 153. We report the first example of an ASM presenting with repeated episodes of aseptic meningitis secondary to an associated intraspinal dermoid tumor. Three other family members of the proband are also known to have ASM.

The use of computerized tomography (CT) in conjunction with metrizamide myelography in the diagnosis of these lesions is also discussed.

Case Report

This 21-year-old woman was referred from another institution. Four years previously, a posterior surgical approach to obliterate her sacral meningocele was unsuccessful because of neural elements entering the meningocele. The patient did well following this operation save for occasional episodes of mild headache and transient leg weakness, which were not serious enough to prompt further medical attention. Three weeks prior to her present admission, however, she experienced the sudden onset of severe headache, fever, lower extremity weakness, and signs of meningeal irritation. A spinal tap revealed cerebrospinal fluid (CSF) with a white blood cell (WBC) count of 2810/cu mm, but no organism was isolated. She recovered uneventfully after 4 days. One week prior to her present admission, the meningitis recurred and CSF obtained by spinal tap at that time demonstrated a WBC count of 483/cu mm, 98% of which were neutrophils. Again, the CSF was sterile and the patient recovered soon after. A CT scan of the abdomen and lumbosacral spine revealed a large ventral sacral mass. The patient had no complaints referable to this mass other than chronic constipation.

The patient’s family history was significant in that her brother had undergone surgical correction of an ASM. Her father and uncle had x-ray evidence of defects, but they had never been symptomatic.

Examination. The patient’s general medical examination and neurological examination were unremarkable. Specifically, sacral sensation was preserved throughout; however, pelvic and rectal examination demonstrated a large firm mass posterior to the rectum. The midline lumbosacral scar from her previous surgery was well healed. There were no cutaneous stigmata. While she was hospitalized prior to surgery, the patient experienced two additional episodes of meningitis. Lumbar puncture after each incident revealed WBC counts of 1500 and 2650/cu mm, with 98% and 93% neutrophils, respectively. The CSF protein was normal on each occasion, and the glucose levels were approximately 35 mg%. All CSF cultures were negative.

Plain x-ray films and polytomograms of the sacrum revealed a large scimitar-shaped defect on the right extending from S-2 to S-4, as well as agenesis of the lower sacral laminae. A barium enema showed the
Anterior sacral meningocele

FIG. 1. Axial computerized tomography scan at the sacral level after instillation of metrizamide into the subarachnoid space demonstrating the anterior sacral meningocele (A). The hypodense intrathecal mass is a dermoid tumor (B).

rectosigmoid region of the colon to be displaced laterally to the right. An intravenous pyelogram demonstrated only lateral displacement of the ureters, with no duplication or other anomalies of the collecting system. Metrizamide myelography confirmed the communication between the thecal sac and the anterior sacral mass, but could not precisely outline the extent of this communication because of dilution of the contrast medium. Computerized tomography of the abdomen and lumbosacral spine immediately after myelography, however, demonstrated the extent of the pelvic mass and its communication with the thecal sac through the anterior sacral defect (Fig. 1). The scan also showed tethering of the spinal cord to a hypodense intraspinal mass which, because of the recurrent aseptic meningitis, was presumed to be a dermoid tumor (Fig. 1). Figure 2 represents an artist’s reconstruction of the pathological anatomy and the relationship between the ASM, dermoid tumor, and tethered spinal cord.

Electromyography revealed only mild paraspinal muscle irritation at L-5 and S-1 on the left. Somatosensory evoked potential testing showed mild changes in the sacral dermatomes bilaterally, slightly worse on the left.

Operation. After the patient had recovered from her last episode of meningitis, surgery was carried out via a posterior transsacral approach. Following a laminectomy from L-4 to S-1 and dissection around the lower dorsal midline sacral defect, a large firm mass could be appreciated within the sacral canal. When the dura was opened, the filum terminale was found attached to the rostral pole of the intraspinal dermoid tumor, thus tethering the spinal cord. The filum was surgically released at its junction with the dermoid tumor, and the tumor was excised. The communication between the thecal sac and ASM was then obliterated. No neural elements were found entering the ASM. The patient remained neurologically intact postoperatively and had no further episodes of aseptic meningitis.

FIG. 2. Artist’s reconstruction of the pathological anatomic relationships.

Discussion

Patients with anterior sacral meningoceles customarily present with symptoms related to the pelvic mass itself, such as chronic constipation, urinary disturbances, and dystocia. Bacterial meningitis is not uncommon following transabdominal surgical approaches or needle aspiration of these lesions. Rarely, ASM may even present with bacterial meningitis. 11,14

Discovery at surgery of a dermoid tumor in conjunction with an ASM is not unique. 5,7,15,16 The preoperative demonstration of such a tumor by CT scanning after intrathecal metrizamide is new, however, and the tumor’s presumed role in the etiology of the aseptic meningitis has not previously been described. The mechanism for the meningitis is thought to be episodic leakage of the contents of the dermoid tumor into the subarachnoid space via a fistula.

Familial association of ASM has been reported previously as a dominant sex-linked trait, 6 and also with no specific gender predominance. 1,10 Males are often asymptomatic, as were two of the relatives alluded to in this case.

The diagnostic work-up should include plain x-ray films of the lumbosacral spine as well as an intravenous pyelogram to rule out associated renal anomalies. A CT scan following metrizamide myelography with computerized reconstruction techniques provides better radiographic evaluation of an ASM than conventional myelography. 4 In this case, it also allowed preoperative diagnosis of an associated dermoid tumor.

Surgical options consist primarily of either a posterior transsacral or an anterior transabdominal ap-
proach. Although Amacher, et al., stated no preference, several recent reports have stressed the advantages of the posterior transsacral technique. It is also evident that the posterior approach affords the best way to deal with possible associated spinal lesions, such as a dermoid tumor and a tethered cord.

References


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