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Abbreviated Reports

Multiple Meningeal Cysts in Marfan Syndrome

Marfan syndrome is an autosomal dominant disorder of type I collagen. Degrees of expression and multisystem involvement, including the eye, heart, and skeleton, are variable [1]. Scoliosis and posterior scalloping of the vertebral bodies commonly are observed [2], and sacral meningoceles have been described [3]. The presence of meningeal cysts in association with Marfan syndrome, however, has not been reported before.

Case Report

A 4-year-old boy with Marfan syndrome (diagnosed when he was 7 months old) had had progressive weakness in the lower extremities for 2 months. A 10-cm mass was palpable in the right lower quadrant of the abdomen. The lower extremities were totally flaccid, with absent deep tendon reflexes but intact sensation. The Babinski sign was not elicited.

Plain radiographs showed generalized cardiomegaly, bilateral posterior mediastinal masses, a soft-tissue mass in the right side of the abdomen, and scoliosis of the thoracolumbar spine with scalloped or concave margins of the lumbar vertebral bodies (Fig. 1). MR imaging of the chest and abdomen showed multiple cystic structures in the posterior mediastinum and retroperitoneum that communicated with an extradural spinal meningeal cyst through intravertebral foramina. Widening of the spinal canal and compression of the spinal cord were present from the lower thoracic spinal canal through the level of L4. The spinal canal widened again in the sacral region, with expanded neural foramina communicating with two presacral cysts. MR clearly showed the spinal and extraspinal meningeal cysts and obviated myelography or contrast-enhanced CT.

Primary resection of the intraspinal cyst was considered but was not performed because of the extent of the cyst and the risk of performing multiple laminectomies in this child who had multiple severe cardiac anomalies. Initial treatment consisted of a lumboperitoneal shunt and drainage of 700 ml from the large retroperitoneal cyst in the right lower quadrant. This resulted in temporary improvement of muscle strength in the lower extremities. The cyst fluid was the same as the lumbar CSF, and multiple samples were clear and colorless with no cells, protein concentrations <20 mg/dl, and glucose levels of 60–100 mg/dl. Marsupialization of the cyst to the peritoneum was performed after the leg paralysis recurred and fluid reaccumulated in the cyst. Pathologic examination of the cyst wall showed

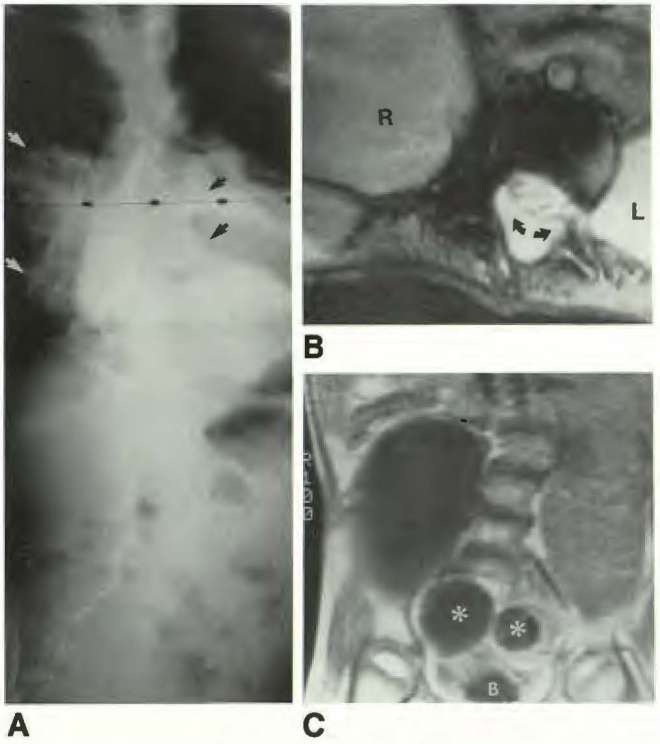

Fig. 1.—Multiple meningeal cysts in Marfan syndrome.

***A*, Anteroposterior spinal radiograph shows bilateral, lobular, posterior, mediastinal masses (*arrows*), cardiomegaly, scoliosis, and a large soft-tissue mass occupying right side of abdomen. Lumbar vertebral bodies have concave lateral margins.**

***B*, Axial T2*-weighted MR image (gradient-recalled echo with 30° flip angle, 500/12) obtained with a surface coil at the L2 level shows bilateral high-signal-intensity retroperitoneal cysts (R and L). Signal intensity diminishes as distance increases from center of surface coil. Posterior intraspinal cyst bulges through intervertebral foramina (*arrows*). Retroperitoneal cyst on left tapers medially toward left intervertebral foramen.**

***C*, Coronal T1-weighted MR image (800/20) of lumbar spine and retroperitoneum shows large low-signal-intensity mass in right retroperitoneum with a tapered margin at L3 level. Image plane is anterior to left retroperitoneal cyst. Two presacral cysts (*asterisks*) and bladder (B) are shown also.**

mesothelium consistent with arachnoid tissue without a covering of dura mater. The patient was able to move his legs against gravity on the first postoperative day, and he walked with assistance within 5 days. Follow-up at 1 year showed no recurrence of paralysis in the lower extremities. The patient could ambulate independently and was continent of stool and urine.

Discussion

Meningeal cysts are diverticula of the spinal meningeal sac, nerve root sheath, or arachnoid that contain CSF. They usually communicate with the subarachnoid space by a narrow stalk, which may act as a check valve, creating an expansile lesion. One criterion for establishing the diagnosis of meningeal cyst has been the histologic identification of a single cell layer of arachnoid without dura mater lining the cyst, but it is common to find only nonspecific connective tissue in the cyst wall [4]. Meningeal cysts may be intradural or extradural and have been found intracranially and intra- and extraspinally. Clinical signs and symptoms are caused by compression of the adjacent brain, spinal cord, or nerve roots [5].

Most meningeal cysts are thought to be congenital, arising in the septum posticum [6] from misplaced nests of arachnoid cells [7], or by herniation of the arachnoid through congenital dura diverticula [8]. Nabors et al. [4] recently proposed a simplified classification of spinal meningeal cysts that includes arachnoid cysts, perineural cysts, and intraspinal meningoceles. They divided the cysts into three categories: spinal extradural meningeal cysts without nerve fibers (type I), spinal extradural meningeal cysts with nerve fibers (type II), and intradural meningeal cysts (type III). This scheme integrates previously used multiple confusing terms into one classification that correlates well with radiologic findings and surgical management.

The thoracolumbar intraspinal cyst and sacral cysts described in our case are type I meningeal cysts. We postulate that the marked dural ectasia present in this child with Marfan syndrome resulted in herniations of the expanding intraspinal meningeal cyst through intervertebral foramina, forming multiple extraspinal cysts. Drainage of the large retroperitoneal cyst resulted in dramatic clinical improvement. Although recurrence of symptoms had not occurred 1 year after marsupialization of the abdominal cyst, septation or blockage of drainage from the intraspinal cyst are possible, because the cyst wall was not excised surgically.

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