Leptomeningeal cysts in congenital ectopia lentis

Case report

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A case of ectopia lentis (Marfan syndrome) with a cerebral subarachnoid cyst, a spinal extradural cyst connected to a lateral intrathoracic meningocele, and perineural cysts is presented. The association of these leptomeningeal cysts in this genetic disorder of connective tissue suggests a common pathogenesis.

Key Words - leptomeningeal cysts - congenital ectopia lentis - Marfan syndrome - spinal extradural cyst - intrathoracic meningocele - perineural cyst

The Marfan syndrome is a disease of mesodermal or ectodermal tissues caused in most cases by a mendelian autosomal abnormality. Degeneration of elastic lamellae appears to be responsible for the characteristic aortic manifestations. The etiology of the skeletal and ocular abnormalities is uncertain but may be a collagen abnormality.

Case Report

This 13-year-old boy was hospitalized for investigation of pulmonary symptoms. Previous illnesses had included multiple episodes of otitis media and pulmonary infection.

Examination. The patient was 5 ft 11 in. tall and weighed 93 lbs. His extremities were long and thin as were his digits. There was a moderate thoracic kyphoscoliosis, ectopia lentis, and abnormal dentition with a high arched palate. A harsh systolic murmur was heard. Neurological examination was normal except for a mild left exotropia. There was no dysphasia, facial weakness or arm drift, reflex asymmetry, or pathological reflexes. Myelography performed with the patient prone disclosed an extradural lesion opposite an area of interpedieular widening and pedicular flattening at T10-11 (Fig. 1 left); this was adjacent to a thoracic mass visible on the plain film (Fig. 1 right); a dilated asymmetrical distal dural tube with perineural sacral cysts was also visualized (Fig. 2). The contrast medium did not enter the extradural or thoracic lesion.

Operation. Thoracic laminectomy exposed an extradural cyst containing clear fluid whose thin wall grossly resembled arachnoid. There was a minute point of connection to the dura at the junction of the dural tube and the dorsal aspect of the T-10 nerve root. The portion of the cyst in the
Leptomeningeal cysts in congenital ectopia lentis

spinal canal measured approximately $3 \times 2 \times 1$ cm. The cyst narrowed and left the spinal canal through the intervertebral foramen at T10-11. Thoracotomy revealed a cyst underlying the parietal pleura; this was easily dissected in the spinal canal and its connection with the dura ligated and cut. The cyst was then pushed through the intervertebral foramen into the thorax where it was dissected out in toto. A pleural flap was sutured over the intravertebral foramen. Both lobes of the lung were emphysematous. An isolated cystic portion of lung, whose only attachment to the lower lobe was by fine strand-like adhesions, was removed.

**Histological Examination.** The wall of the cyst consisted mainly of collagenous fibers with what appeared to be interspersed flattened arachnoidal cells.

**Postoperative Course.** Approximately 1 year later the patient was admitted to another hospital with signs of heart failure. Angiography revealed dilation of the aorta. Conservative therapy failed, and he died during aortic surgery.

**Postmortem Examination.** Autopsy revealed cystic necrosis of the media of the aorta and, to a lesser extent, the pulmonary arteries. There was severe aortic incompetence plus myocardial hypertrophy and evidence of congestive failure in the liver and lungs. There was also mesangial hyperplasia in the renal glomeruli and numerous subcapsular areas of lymphangiectasis within the spleen. An incidental finding was a large arachnoid cyst displacing the frontal and temporal cerebral lobes on the left (Fig. 3). The spinal cord was not examined.

**Discussion**

Lombardi and Morello have proposed that leptomeningeal intradural cysts, spinal extradural cysts, and perineural cysts are the
result of common developmental factors and differ in evolution and symptomatology according to their location. Those located in the telencephalon would be classified as arachnoid cysts and those in the spine as intra- or extradural arachnoid cysts, lateral intrathoracic meningoceles, anterior sacral meningoceles, lumbosacral posterior meningoceles, or perineural cysts.

Primary subdural arachnoid cysts are rare. Extrudal cysts are usually of congenital origin and are presumably protrusions of arachnoid through congenitally weakened places in the dura; they may or may not retain a patent subarachnoid space at the point of connection through the dura. Approximately half the reported cases of extradural arachnoid cysts have been associated with dorsalis kyphosis juvenilis. Extrudal cyst walls are composed of flattened cells and laminated collagenous walls.

About 70% of the cases of lateral intrathoracic meningoceles have been associated with von Recklinghausen’s disease, which is also an autosomal dominant genetic disorder. The associated bone changes frequently include kyphoscoliosis, dorsal excavations of the vertebral body, and enlargement of the intervertebral foramina, but the cortical margins of the canal and vertebral bodies are usually normal. These are similar to the bone abnormalities seen in the Marfan syndrome. In von Recklinghausen’s disease the pathogenesis of the meningoceles probably involves both a primary dural malformation and degeneration and associated bone changes. Pathologically, the lateral intrathoracic meningoceles are considered to be protrusions of the dura through enlarged intervertebral foramina. In our case, however, the lining of the meningocele clearly did not resemble dura; it was quite transparent and grossly was identical to arachnoid.

The origin of perineural cysts is unclear. Tarlov considered them to be the result of either ischemic degeneration in ganglia or subarachnoid hemorrhage distending perineural spaces. He felt that the sacral cysts did not communicate with the subarachnoid space. However, myelography makes it quite clear that in many cases there is in fact a communication. They may result from a
Leptomeningeal cysts in congenital ectopia lentis

developmental abnormality consisting of arachnoidal proliferation and subsequent cavitation. More recently it has been suggested that they develop at the point of least resistance to the hydraulic pressure of CSF operating to dilate the epineurium. Strully and Smith also support the view that the cyst-like formations are meningeal diverticulae produced by hydrostatic pressure transmitted through the subarachnoid space. They base this theory on the clinical fact that there is usually no history of previous trauma or inflammation; the anatomical evidence includes the symmetry of the nerve root involvement, the greater incidence and size of caudally located cysts, and the fact that the spinal subarachnoid space does extend out along the dorsal nerve root sleeve to the ganglion.

Nelson noted enlargement of the spinal canal involving both depth and width, narrowing of the pedicles, widening of the interpedicular distances, and posterior scalloping of the vertebral bodies. His four patients did not have contrast studies to delineate intraspinal pathology, which he implies does not occur in the Marfan syndrome. The developmental enlargement of the spinal canal occurring in the absence of intraspinal lesions in this syndrome has also been documented by Shapiro. Seven of 40 patients with the Marfan syndrome in one review had suffered periodically from low-back pain of sufficient intensity to cause them to seek medical attention. In another case involving a pregnant patient, what was probably an anterior sacral meningocele interfered with delivery.

The pathogenesis of cerebral arachnoid cysts is unclear but it is generally assumed that some of them are primarily developmental. Starkman, et al., have suggested that abnormalities in the usual mechanism by which the leptomeninges are derived from the perimedullary mesenchyma create false passages that may be enlarged by CSF pressure waves.

The embryonic layer of origin for the leptomeninges is not definitely established. Flexner and most authorities have suggested that the meninges, including the pia mater, are of mesodermal origin, but more recent workers have attributed an ectodermal origin to the cells of the inner part of the pia although agreeing that the arachnoid and dura are of mesodermal origin. If the Marfan syndrome is in fact a mesodermal dystrophy, this would tend to support the view that the arachnoid is of mesodermal origin.

Our case with its multiplicity of leptomeningeal cyst types suggests that a common defect in elastic and collagen fibers may have predisposed this patient to the development of intra- and extradural arachnoid proliferations and herniae with unique characteristics resulting from their specific anatomical location.

References

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