Posterior spinal cord herniation into an extradural thoracic arachnoid cyst: surgical treatment

Case report and review of the literature

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The authors describe the case of a 2-year-old boy who experienced progressive spastic paraparesis for several months. Magnetic resonance imaging revealed an extensive extradural arachnoid cyst at the T3–L1 levels and posterior spinal cord herniation at T3–4. Surgical release of the neck of the hernia and total resection of the arachnoid cyst were performed. The patient had good clinical recovery several weeks after surgical decompression. This case highlights an exceedingly rare type of spinal cord herniation in a pediatric age group, and focuses on the abnormalities of the dorsal dura mater, together with imaging and intraoperative findings.

Key Words • extradural arachnoid cyst • spinal cord herniation • paraplegia • pediatric neurosurgery

Since Wortzman, et al., reported the first case of spinal cord herniation, to our knowledge nearly 80 other cases have been reported. Precise descriptions of radiographic and intraoperative findings of spinal cord herniation remain, however, insufficient. This applies particularly to the ventral portion of the dura mater, where the presence of a dural defect situated on the concavity of the spinal curvature is a prerequisite for this rare condition. In this first reported case of the disease in a young child, we describe a patient with idiopathic spinal cord herniation in the upper thoracic area.

Case Report

History and Presentation. This 2-year-old boy presented with progressive paraparesis. The boy had been delivered by an elective cesarian section after his mother had experienced an uneventful pregnancy and 13 years of infertility. There was no history of trauma or infection. He could stand at the age of 9 months and walked without help at the age of 1 year. After several months, progressive paraparesis and gait deterioration developed.

Examination. A neurological examination revealed severe paraparesis and an inability to stand and walk, with increased deep tendon reflexes in the lower extremities. His upper limbs and sphincter function were normal. Radiographs of the thoracic spine showed widening of the spinal canal with thinning of the pedicles at the T3–12 levels. Spinal MR imaging revealed an extradural cyst posterior to the cord that extended from T-3 to L-1. The cyst had several septations and contained fluid that had a similar signal intensity as cerebrospinal fluid. The MR image showed a kink in the spinal cord at the T3–4 level. An arachnoid cyst at those levels seemed to be drawing the cord into its mouth, where a small cord hernia could be seen. At other levels, the dural sac was markedly displaced in a forward direction by a posterior arachnoid cyst (Fig. 1).

Operation. The boy underwent an osteoplastic T3–L1 laminotomy. A thoracic extradural cyst with septations was found that separated the cavities incompletely. In the upper part of the arachnoid cyst, the cord and a root on its right lateral side were herniated into the arachnoid cyst. The herniated cord was pale and appeared as a knob of tissue 20 mm long and 10 mm in diameter. The herniation orifice which was obviously smaller than the cord diameter, was divided to let the cord fall away. The dura was left open widely, and was closed by patch graft around the level of the herniation. The cyst was excised completely.

Postoperative Course. The boy’s postoperative recovery was uneventful, and his paraparesis gradually improved during the course of a month. Two years later, his gait was normal and spinal MR images showed a normal cord signal at the level of the herniation.

Discussion

Spinal cord herniation is very rare and can be classified...
Posterior spinal cord herniation

based on its origin as idiopathic, posttraumatic, or iatrogenic. As Martin \(^4\) has mentioned, it occurs most commonly ventrally, in the midthoracic region, because the thoracic curvature tends to pull the spinal cord anteriorly. Some authors have postulated that the spinal cord herniates into an extradural arachnoid cyst, \(^3,8\) whereas others report that it herniates into a cavity produced by congenital duplication of the dura. \(^1\) More than 90% of spinal cord herniations occur in patients who are at least 30 years old. \(^2\) The possibility of thoracic disc herniation as a cause of this dural defect was reviewed by Miyaguchi, et al., \(^6\) and a degenerative process with slow erosion of the dura would seem consistent with the age at presentation. \(^5\) Although herniation usually occurs only later in life, the fundamental pathology per se may be congenital.

The boy in this case is the youngest patient reported so far with neurological deficits due to spinal cord herniation. Posterior spinal cord herniation is rare, as is its association with an arachnoid cyst. This is the second report of such a case. This posterior impaction may involve a lower pressure within the arachnoid cyst or a higher pressure in the canal, as would be generated by a Valsalva maneuver. In such a situation, adhesions develop between the cord and the edges of the dural defect, and cerebrospinal fluid pulsation pushes the cord into a preexisting cyst.

The patient reported on here had no history of trauma, infection, or surgery. The arachnoid cyst and subsequent cord herniation into the dural sac were most probably congenital. Symptoms such as these probably occur when herniation fills the orifice and strangulation takes place, which explains the late appearance and progressive evolution of this myelopathy. Most of the patients in the reported cases of anterior spinal cord herniations presented with symptoms of Brown–Sécuard syndrome. \(^7\) Martin \(^4\) and we both describe a patient who presented with progressive paraparesis—a symptom different from that associated with anterior cord herniation.

Conclusions

Analysis of the reported cases leads to the conclusion that without treatment the condition is usually progressive, and paraplegia is a possible outcome. The proposed treatments are to reduce the hernia by incising the neck and opening the membranes widely to prevent impaction. Mobilization of the herniated spinal cord into the canal is achieved by surgery that can stop the evolution of symptoms and signs.

References


FIG. 1. Sagittal T2-weighted MR image showing an extensive extradural arachnoid cyst with cord compression and posterior cord herniation at the T3–4 levels.