Neuroenteric Cyst: A Rare Cause of Childhood Paraplegia

Neuroenteric cysts are rare congenital anomalies derived from the displaced endodermal tissue around the third week of the embryonic stage. We report a case of neuroenteric cyst of the spinal canal presenting with paraplegia in a 2.5-year-old boy. Despite the 12-day delay in surgical decompression, he made a complete neurological recovery. This is a unique case in regard to the clinical presentation, MRI findings and the excellent outcome after surgery.

Keywords: Neuroenteric Cyst, Paraplegia, Magnetic Resonance Imaging

Introduction

Neuroenteric cysts (NC), also called enterogenous cysts, are rare anomalies of the central nervous system. They result from abnormal separation of the neural tube and the endodermis during the 3rd week of gestation, leading to the persistence of endodermal elements in the spinal canal.\(^1\)\(^-\)\(^3\) They are sometimes found only in the mediastinum, and associated vertebral anomalies are frequent.\(^4\)

They represent approximately 0.7% of the tumors and 16% of the cysts in the CNS. Five percent of the patients with Klippel-Feil syndrome and vertebral fusion abnormalities may have enteric cysts.\(^5\)

The clinical presentation of NC is usually insidious, consisting of symptoms and signs of progressive myelopathy.\(^6\) Decompressive surgery is the treatment of choice. The outcome is usually good\(^1\)\(^,\)\(^6\) because of the mild preoperative symptoms. We report a case of an intraspinal thoracic NC that presented as acute paraplegia. A full neurological recovery was achieved with surgical decompression despite a 12-day delay in treatment.

Case Presentation

A 2.5-year-old boy presented with a four week history of abdominal pain and urinary incontinence about 10 days before admission. He got medication for suspected pyelonephritis and consequent improvement in his symptoms as a result. Kidney ultrasound at the time of infection was normal.

Three days later, the movements of his legs were apparently weak and he was not able to walk. Therefore, the child was referred to the neurosurgical department. There was no history of fever, altered consciousness or seizure.

On admission, movements and sensation of the lower limbs were severely affected and he developed urinary retention. He had no familial or underlying diseases.

The summary of routine plasma biochemistry and CSF analysis is shown in Table 1.

The neurological examination revealed decreased muscular tone of both feet,
diminished deep tendon reflexes and positive plantar responses. He had flaccid paraplegia and a sensory deficit below the D4 dermatome and also bladder retention.

On investigation with 0.5 Tesla GE MRI, with and without contrast revealed evidence of a non-enhancing well-defined intradural extramedullary cystic lesion extending from D2 to D6 of the vertebral level, situated posterior to the cord causing anterior displacement with marked cord compression at this level (Fig.1). Complete fusion anomaly at D4-D5 level are seen in Figure 1.

Pathologic Findings

The cyst contained fluid that was slightly thicker than the cerebrospinal fluid.

Biopsy of the cyst wall revealed pseudostratified columnar epithelium lining the cyst wall. Some of the cells were ciliated and some had secretory (goblet cells) changes consistent with neuroenteric cyst, for-gut (bronchogenic) cyst (Fig. 2).

Discussion

Definition and Origin

NCs are rare congenital cysts of the spinal canal lined with epithelium of endodermal origin. Controversy persists about their exact embryopathogenesis, but it is well established that they result from a disruption occurring in the 3rd week of embryogenesis and involving the complex series of interactions responsible for the formation of the notochord, neuroenteric canal, endoderm, and neural tube.2

Location and Associated Anomalies

NCs are usually intraspinal, with an increased frequency at the cervicothoracic junction and in the inferior cervical region.1 The cysts are most frequently anterior to the spinal cord, with only a few reported posterior to it.8 They can be extradural or intradural, and occasionally intramedullary.9 They are frequently associated with vertebral anomalies: hemivertebra, absence of the vertebra, anterior and posterior spina bifida and kyphosis.4,10 They can also be associated

<table>
<thead>
<tr>
<th>Table-1: Laboratory Test Result</th>
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<tr>
<td>WBC: 7700/mm³</td>
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<tr>
<td>Hb: 13.1 mg/dl</td>
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<tr>
<td>Hct: 38.6%</td>
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<tr>
<td>MCV: 82.3 fl</td>
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<tr>
<td>MCH: 27.9 pg</td>
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<tr>
<td>MCHC: 34.3%</td>
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<td>Platelet: 361000/mm³</td>
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<tr>
<td>LDH: 1034 U/L</td>
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<tr>
<td>U/A: NORMAL</td>
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<tr>
<td>U/C: NEGATIVE</td>
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<tr>
<td>CA: 9.8 mg/dl</td>
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<tr>
<td>CSF analysis:</td>
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<tr>
<td>Appearance: clear</td>
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<tr>
<td>Protein: 140 mg/dl</td>
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<td>Culture: negative</td>
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Fig. 1. 2.5-year-old boy with neuroenteric cyst; A. T1 and T2 Weighted sagittal MRI images with contrast revealed evidence of a non-enhancing well-defined intradural extramedullary cystic lesion extending from D2 to D6 of the vertebral level, situated posterior to the cord causing anterior displacement with marked cord compression at this level. B. T1 and T2 Weighted axial MRI images with contrast revealed evidence of a non-enhancing well-defined intradural extramedullary cystic lesion extending from D2 to D6 of the vertebral level, situated posterior to the cord causing anterior displacement with marked cord compression at this level.
with diastematomyelia or syringomyelia. Intrathoracic or intra-abdominal cysts can coexist with NCs, with a fibrous band joining them to the intraspinal NC through a vertebral defect. An open communication between the two cysts is rare. In the case presented here, the NC was in the typical site at the thoracic region, but in an unusual location, posterior to the spinal cord, with vertebral fusion anomaly.

Clinical Manifestations and Diagnosis

Wilkins et al. reviewed 119 patients with an intraspinal NC (72 men and 43 women). The diagnosis of NC was established during the first decade of life in 34% of cases and in the second decade in 23% of cases. The clinical spectrum of NC depends on the site of the lesion and includes local pain, radiculopathy and/or myelopathy. Neonates and young children frequently present with symptoms related to an intrathoracic or intra-abdominal cyst; they rarely lead to a picture suggestive of meningitis. The onset of symptoms is generally insidious, and progression takes place over a prolonged period.

Neurological signs can be conspicuously minimal or absent despite severe compression of the spinal cord. Progressive clinical manifestations may follow trauma. The possible explanations for this unusual presentation are sudden mechanical compression of a chronically distorted and compressed spinal cord, or an increase in the size of the cyst as the result of an accumulation of intra-cystic fluid. One case of a subacute presentation of NC was reported by Derlon et al.: a spinal hemiplegia appeared over the course of several days. Our case is unique in its clinical manifestations: urinary tract infection and urinary incontinence followed by acute paraplegia that can be considered as traumatic or an increase in the size of the cyst as the result of an accumulation of intracystic fluid.

Differential diagnosis includes arachnoid and neuroepithelial cysts, epidermoid cysts, cystic schwannoma and inflammatory cysts such as cysticercosis. MRI is the diagnostic test of choice for NC. NC has an iso to slightly hyperintense signal relative to that of CSF on T1W and T2W images. The MRI findings in our case were consistent with those accepted for diagnosis.

Treatment

NC should be treated by decompression and as near-complete excision of its membrane as possible. However, complete resection of the cyst wall is not always possible because of the presence of tight adhesions to the spinal cord. In these cases, subtotal excision results in a benign course, and most authors recommend it in preference to aggressive complete removal of the lesion, which carries the risk of heavy neurological lesions. The optimal surgical approach remains controversial. The anterior approach is indicated for several reasons; complete resection is easier because the cyst is generally anterior to the spinal cord, and there is less risk of trauma to the spinal cord and of kyphosis. In addition, better management
of cases associated with complex vertebral anomalies is possible. While these concerns are valid, they remain theoretical; the posterior approach assures a good outcome, is technically simpler, and remains the preferred one in most instances. Subtotal resection of the cyst was performed because of tight adhesions to the anterior. Pathologic examination usually reveals a cyst lined with pseudostratified, simple cuboidal, or columnar epithelium, which can be ciliated. This epithelium may show gastrointestinal or pancreatic elements. Biopsy of the cyst wall revealed pseudostratified columnar epithelium lining the cyst wall. Some of the cells were ciliated and some had secretory (goblet cells) changes consistent with neuroenteric cyst, forgut (bronchogenic) cyst.

Outcome

The long term neurological outcome of NC treatment is good, especially in children. This is due to moderate preoperative neurological signs. Despite the 12-day total paraplegia, our patient was able to stand 3 months after surgery, showing a near complete neurological recovery. This is very rare in the literature and is observed only when severe compression of the spinal cord has lasted for several days. It has been suggested that children with spinal cord compression might have a much better prognosis for recovery of neurological function than adults. Recurrences of NC can be observed, especially after partial excision. However, total resection of the cyst is not always possible, as in our case, because of tight adhesions to the spinal cord.

One year later he was able to walk and was continent for urine and feces.

References