Case Report

Changes in Cervical Neurenteric Cyst Size

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Keywords

Cervical spine · Magnetic resonance imaging · Spinal cyst · Neurenteric cyst · Pediatrics

Abstract

Spinal neurenteric cysts are rare congenital anomalies and the natural history of the cyst is not fully understood. We evaluated a case of spontaneous absorption of a cyst. The patient was a 5-year-old boy who had experienced pain in the bilateral upper extremities 3 days before his admission. Neurological examination revealed severe motor weakness of the upper extremities and mild motor weakness of the lower extremities. Magnetic resonance imaging (MRI) revealed an extramedullary cystic lesion with a fluid-fluid level in the cervical region. One week later, the pain spontaneously improved with bed rest, and the patient showed good neurological recovery. MRI performed 6 months later revealed spontaneous reduction of the cyst. Enlargement and contraction of the cyst were observed over 3 years on MRI. Since the size of the cyst changes, MRI should be performed annually for patient follow-up.

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Introduction

Neurenteric cysts are uncommon congenital lesions, and their pathogenesis is unknown. At present, it is largely accepted that the cysts are a form of split notochord malformation, consisting of a cyst lined by simple or pseudostratified columnar cells [1, 2]. In rare cases, especially in children, spontaneous rupture of the cyst has been observed [3]. However, the natural history of such cysts is still unknown, and only a few reports have described cases of spontaneous absorption on imaging studies, such as magnetic resonance imaging (MRI).

We herein report the clinical and radiographic details of a case of spontaneous cyst absorption in a 5-year-old boy.

Case Report

A 5-year-old boy born to nonconsanguineous parents with no history of any major illness presented with sudden severe neck pain. Three days after the onset of the symptom, severe radicular pain developed in both upper limbs followed by sudden weakness in both lower limbs, prompting presentation to the emergency department. Upper limb neural tension tests were not possible due to his upper limb pain. Neurological examination revealed motor deficit in the upper and lower limbs (motor strength: 2/5 in the upper limbs and 4/5 in the lower limbs), and the symptoms progressed to difficulty in walking. Light touch and pinprick sensations were decreased over both hands. Normal deep tendon reflexes were noted in the upper and lower extremities, and Babinski reflex was negative. He could urinate without difficulty. The Japanese Orthopedic Association score was 6 points (total: 17 points).

MRI of the cervical spine revealed an extramedullary cystic lesion extending to the C5 level, causing expansion of the spinal cord. The mass was isointense to the spinal cord on T1-weighted imaging, hyperintense on T2-weighted imaging, and showed faint peripheral enhancement with the administration of gadolinium. The lesion showed a well-defined fluid-fluid level and was located at the anterior part of the extramedullary region (Fig. 1). There were no associated bony or soft tissue abnormalities.

We planned an operation to remove the cyst with an anterior approach involving vertebrotomy; however, after 3 days of bed rest, the patient's cervical and upper limb pain had completely improved, and the bilateral hand numbness was significantly improved. He had 4/5 power in his bilateral grip and full strength otherwise. Three weeks later there was no change in tumor size on MRI, but the patient was able to walk without support. The patient's parents were provided with a detailed explanation of the benefits, limitations, and possible risks of surgery for the purpose of obtaining their informed consent. However, they refused surgery because of the dramatic improvement in the patient's symptoms. MRI 6 months later revealed spontaneous reduction of the cyst with no evidence of neural compression (Fig. 2a). MRI was performed regularly for the next 3 years, and the cyst showed a cycle of shrinking and enlarging (Fig. 2b–d). However, there was no recurrence of the symptoms in the 3 years, and the patient currently has no problems in his daily life. His Japanese Orthopedic Association score is 17 points.

Discussion

Spinal neurenteric cysts are rare, accounting for 0.3–1.3% of all spinal tumors [2]. Neurenteric cysts are 1.5–3 times more common in males than in females, and most are discovered in infancy [4]. Although several theories have been proposed, the pathogenesis is still unknown. There is one theory that accounts well for the formation of the neurenteric canal during embryogenesis. During the third week of gestation, the neuroectoderm may fail to separate from the endoderm, leading to the growth of endodermal cells within the neuraxis and thus development of a neurenteric cyst [1, 2, 5]. Spinal neurenteric cysts are usually cervicothoracic in their location and are often ventrally situated [1]. In the past, the diagnosis was usually made during surgery. The differential diagnosis includes subarachnoid cyst, cystic dermoid, epidermoid and teratoma cyst, parasitic cyst, and ependymal cyst, which may resemble neurenteric cysts. Cystic teratoma may be very difficult to distinguish from neurenteric cyst and can show similar histopathological characteristics. Neurenteric cysts are typically located ventral to the spinal neuraxis, whereas teratomas are usually located dorsal to the spinal cord and are not associated with other congenital malformations. The location of the cysts can be crucial, and MRI has become the most important diagnostic tool for these lesions [5]. Lack of avid contrast enhancement and absence of a mural nodule distinguish neurenteric cysts from other spinal tumors [6]. With the availability of MRI, the diagnosis can now be made preoperatively. In our patient, no pathological material was available for review; however, on MRI, classic findings of neurenteric cyst were observed. These findings were considered sufficiently characteristic for a definitive diagnosis.

Patients harboring a spinal neurenteric cyst may present with pain, myelopathy, progressive weakness, or sensory dysfunction [4]. Most neurenteric cysts are benign and slow-growing, so symptoms tend to be insidious at onset. In rare cases, especially in children, spontaneous rupture of the cyst and spillage of its contents into the subarachnoid space can cause a meningitis-like condition [3, 4]. Spillage of caustic cyst contents into the subarachnoid space might have caused the inflammation of the neural elements and pain in the present patient. Reports exist, however, of intermittent symptomatology and sudden neurological deterioration, which are both believed to occur due to leakage of the cyst contents [2]. In some patients, the clinical course is characterized by exacerbations and remissions, which are attributed to periodic rupture of the cyst contents or changes in the rates of mucin production and reabsorption by the cyst wall [7]. Our patient presented with sudden onset of symptoms; however, his pain improved spontaneously, and he showed good neurological recovery. MRI revealed spontaneous reduction of the cyst, and enlargement and reduction of the cyst were observed on MRI during 3 years of follow-up.

Most neurenteric cysts are extramedullary, and there is a clear plane of dissection between the lesion and the neural elements. Simple aspiration is unacceptable because it generally results in recurrence of the cyst. Since neurenteric cysts are benign, resection is the treatment of choice. The goal of the initial surgical intervention is resection of both the cystic contents and the cyst wall [3]. Our patient is still a child, and we are performing follow-up because of the possibility that surgery would be associated with development of cervical spine deformity and difficulty of spinal fusion. There is a risk of recurrence following spontaneous reduction; thus, it is necessary to continue follow-up with MRI for these patients.

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Statement of Ethics

The patient and his parents were informed that the data from his case would be submitted for publication and consented to this; the parents provided written consent.

Disclosure Statement

The authors declare no conflicts of interest in association with the present study.

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Fig. 1. Magnetic resonance images of the patient's spine. **a, b** Sagittal (**a**) and axial (**b**) T2-weighted images showing a hyperintense signal in the extramedullary region. **c, d** Sagittal (**c**) and axial (**d**) gadolinium-enhanced images showing peripheral enhancement. The images show a cystic lesion causing significant spinal cord compression.

Fig. 2. Sagittal T2-weighted magnetic resonance images of the patient's spine. **a** Images obtained 6 months after the first visit demonstrating cyst shrinkage. The cyst could barely be confirmed. **b** Enlargement of the cyst was visible on the image obtained 1 year after the first visit. **c** Imaging obtained 2 years after the first visit demonstrating cyst shrinkage. **d** Enlargement of the cyst was visible on the image obtained 3 years after the first visit.