Neurenteric cysts are rare malformations that lead to brain stem and spinal cord compression. These cysts are derived from endoderm that is fused with the developing notochord during the third week of gestation. A 3-year-old boy presented with a 2-month history of torticollis and had developed paraparesis 2 weeks before admission. Examination showed that he was quadriparietic. Plain radiography films revealed widening of the cervical canal and pedicle erosion, while a hypointense lesion on T1, at the craniovertebral junction, with a compression effect on the anterior aspect of the brain stem and spinal cord were noted on magnetic resonance imaging. C1 to C3 laminectomy as well as a suboccipital craniectomy were performed; upon opening the dura, a cystic swelling was found to be protruding. Fluid was aspirated from the cyst and after shrinkage, the cyst was removed completely. On the first day after the surgery, the patient’s motor power improved significantly. This report describes a rare form of neurenteric cyst in terms of both its location and unusual presentation (torticollis).

Keywords: central nervous cyst, endoderm, entogenous cyst, neurenteric cyst, torticollis

Introduction

Neurenteric cysts are rare congenital anomalies usually presenting as a posterior mediastinal mass. The occurrence of neuroentric cyst at the craniovertebral junction of spinal cord is a very rare phenomenon. The nomenclature of such a lesion, which varies from author to author, includes the terms “teratomatous cyst” and “enterogenous cyst”, probably reflecting the uncertainty of the etiology. This report describes a case of neurenteric cyst of the craniovertebral junction and reviews the clinical aspects of this entity.

Case Report

History

A 3-year-old boy was referred to our clinic because of a 2-month history of torticollis. He had developed paraparesis 2 weeks and urinary incontinence 1 week before the admission. On plain radiography, widening of the cervical canal and pedicle erosion, while a hypointense lesion on T1, at the craniovertebral junction, with a compression effect on the anterior aspect of the brain stem and spinal cord were noted on magnetic resonance imaging (Figure 1). His motor weakness had progressed towards the upper extremities, particularly the right upper limb. The symptoms became progressively worse and led to development of ataxia.

Examination

The patient looked well. The right upper extremity was paretic with Grade I muscle power (medical research council [MRC] scale for grading muscle strength, 1944). The left upper limb and both lower limbs, however, were paretic with Grade III muscle power. Anal sphincter was impaired and beside a positive Babinski’s sign, the reflexes were brisk bilaterally.

He was unable to sit, stand or walk because of severe spastic quadriplegia. A general sensory impairment including loss of pinprick sensation up to the level of the upper cervical region was found. No spinal tenderness was noted.

On magnetic resonance imaging (MRI), a
A hypointense lesion in T1 was observed at the craniovertebral junction, with a compressive effect on the anterior aspect of the brain stem and spinal cord (Figure 1). On plain radiography, widening of the cervical canal and pedicle erosion were seen (Figure 2).

**Operation**

C1 to C2 laminectomy as well as a suboccipital craniectomy was performed. Dura was observed as bulging and soft. Upon opening the dura, a cystic swelling was found to be protruding. Fluid was aspirated from the cyst (Figure 3). After shrinkage, the cyst was removed completely. On the first postoperative day, the patient’s motor power improved significantly. He was discharged 10 days after the operation with normal motor function and no torticollis.

A cystic lesion measuring about 3.5 × 3.5 × 1.5 centimeters was reported by the pathologist. Microscopically, the cyst wall was composed of an outer coat of collagen, with supporting basement membrane, and a layer of cuboidal epithelium contained positive periodic acid-Schiff mucin (Figure 4).

**Discussion**

Neurenteric cysts are rare malformations that lead to spinal cord compression or tethering. The name “enterogenous cyst” was first applied by Harrimon in 1958 to intraspinal cysts, usually extramedullary lined by columnar epithelium of intestinal character.

The possible developmental mechanisms of these lesions have been further discussed by several observers. The review by Agnoli, Laun and Schonmayr lists 33 reported cases. The great majority of these cases were intra- or subdural in nature, but four intramedullary cases were claimed to be related to the mediastinal cysts of similar character, which are associated with developmental defects of the vertebra. According to this hypothesis, defects would permit the
formation of a fistula in early embryonic life, between formation of the neuroectoderm and the yolk sac. However, vertebral anomalies are demonstrable in a minority of cases—only in nine of the 33 reviewed by Agnoli and colleagues.

Therefore, when such anomalies are absent, the mechanism underlying the development of these cysts is largely conjectural. Based on a maldevelopmental factor, enterogenous cysts should be distinguished from the cystic teratoma that may arise in the spinal canal.\(^1\)

Embryogenesis of enterogenous cysts is a matter of debate. Rhaney and Barclay suggested that intraspinal enterogenous cyst was a product of abnormal separation of germinal germ cells.\(^{10, 11}\)

Early in embryonic development, the primary mesoderm, which gives rise to the notochord, comes to lie in close contact with the endoderm, a process called “intercalation”. At a later stage, when separation occurs (“excalation”), groups of endodermal cells may be carried back with the mesoderm to give rise to the enterogenous cyst.\(^{10, 11}\)

Neurenteric cysts can present at a wide range of ages, from the newborn period to the fifth decade of life. They are two to three times more prevalent in males. Malformations of the split notochord syndrome have been diagnosed prenatally by ultrasound and are apparent at birth. The complex association of intestinal protrusion, cloacal/ bladder extrophy, and renal dysgenesis

Figure 2. Frontal view of the cervical spine demonstrating widening of the interpediculate distance in the upper cervical spine.

Figure 3. Intraoperative microscopic views of the neurenteric cyst (C). S = spinal cord.
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may be fatal. The presentation of intraspinal cysts is more common during adulthood. The symptoms of spinal cord or nerve root compression may mimic other space-occupying lesions of the spinal canal, including disc herniation. Pain is the most common symptom and localizes to the spinal level of malformation.

Bacterial meningitis can occur when a neurenteric cyst is accompanied by dorsal sinus tract, whereas inflammation of the cerebrospinal fluid from contact with cyst contents is unusual.

In this report, a rare form of neurenteric cyst located in the craniovertebral junction with an unusual presentation of torticollis was described.

To our knowledge, this is the first report of clinical presentation of neurenteric cyst as a torticollis. The patient’s motor function improved significantly and torticollis disappeared on the first postoperative day following laminectomy and suboccipital craniectomy.

References