Neurenteric cyst of the cervicothoracic junction: A rare cause of paraparesis in a paediatric patient

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ABSTRACT

Neurenteric cysts are rare congenital spinal masses that result from the dysgenesis of the endoderm tissue during development. We report a 4-year-old girl who presented with an insidious onset of lower limb paraparesis. An MRI scan revealed a cervicothoracic intradural extramedullary neurenteric cyst at the thoracic T1/T2 level, with marked spinal cord compression. No associated spinal dysraphism was noted. The patient underwent laminotomy and excision of the cyst. She recovered her neurological functions completely post-operatively, and at her six-month follow-up she was asymptomatic without any neurological deficits. We will discuss the pathogenesis, clinical presentation, and neuroradiological findings. We emphasize the value of early surgical intervention and long-term follow-up when this type of lesion is only partially excised.

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1. Introduction

A neurenteric cyst is a rare developmental anomaly. It is usually found in the posterior mediastinum and abdominal cavity, and rarely involves the spinal canal.1 An isolated intradural extramedullary cyst is extremely rare, and is usually associated with anomalies of the vertebra. The cysts seen in the spinal canal at the craniocervical junction are mostly extramedullary and occasionally intramedullary. We report on a 4-year-old girl with an intraspinal neurenteric cyst who presented with progressive paraparesis.

2. Case report

A 4-year-old girl presented with a six-week history of difficulty in walking, preferring instead to sit or lie down. With support, she walked with a stooped posture, dragging her left foot and maintaining her neck in a flexed position. She had a power of grade 3/5 in both lower limbs with associated hyperreflexia. Her upper limbs were normal. There did not appear to be a sensory deficit, and the bladder and bowel functions were normal.

An MRI scan of the cervical spine showed an unusual intradural extramedullary mass lesion extending from the C7/T1 junction to the T2/T3 disc space (Fig. 1a). This mass was situated anterior to the spinal cord, causing its posterior displacement and marked compression (Fig. 2). The mass was hypointense on T1-weighted and hyperintense on T2-weighted acquisition images (Fig. 1b). The cyst wall did not enhance following the administration of gadolinium diethylentriaminepenta-acetic acid (Gd-DTPA). No spinal dysraphism or other lesions were identified.

The patient underwent a C7 and T1 laminotomy. The dura was opened and the cyst was identified lying anterior to the spinal cord, compressing and pushing it dorsally. The cyst was drained and the wall was totally excised. The patient's lower limb weakness improved gradually over the next six days, and she was able to walk without support. At her six-month follow-up she was walking with a normal gait, and her lower limb paraparesis had resolved completely.

Histopathological examination of the tissue demonstrated a cyst wall lined with cuboidal to columnar epithelium, with some cells appearing ciliated (Fig. 3). No mitosis was seen.

3. Discussion

Intraspinal neurenteric cysts are rare endodermal developmental lesions, and represent 0.3% to 1.3% of the entire spinal canal tumours.1 The pathogenesis of the neurenteric cyst remains unclear. Dysgenesis of the endoderm with neurenteric canal formation at the notochord in the third week of embryogenesis has been proposed as an explanation of neurenteric cyst formation. The main theories for neurenteric cyst formation include: (i) a primary adhesion of endoderm anterior to the notochord; (ii) incomplete escalation of the notochord; (iii) persistence of the neurenteric canal or formation of an accessory neurenteric canal with a split notochord, which may explain the coexistence of vertebral and gastrointestinal anomalies; and (iv) displacement of endodermal cells.2 Most reports describe neurenteric cysts in the cervical and cervicothoracic junctions of the spinal canal, as with our patient, but these cysts have also been described as occurring in other areas of the spinal canal, such as the conus medullaris. These are rare tumours that develop at the craniocervical and cervicothoracic junctions, with fewer than 20 cases reported worldwide.

Depending on the site of the lesion, intermittent symptoms of myelopathy and radiculopathy are the usual clinical presentations. Neck pain with occipital headache is the most common clinical presentation of a cranio cervical neurenteric cyst. Symptoms such as cranial nerve dysfunction, hemiparesis, paraparesis (as reported in this patient) and meningitis as a result of chemical irritation secondary to cyst fluid leakage or bacteria meningitis have also been reported; burning dysesthesia may occur occasionally.

It is usually impossible pre-operatively to definitively diagnose a neurenteric cyst. MRI has been the main modality of investigation, as for other spinal cord lesions. However, when viewed on MRI, a neurenteric cyst is often mistakenly diagnosed as an arachnoid cyst. However, congenital intradural extramedullary arachnoid cysts are virtually always posterior to the cord. The predilection of a neurenteric cyst to be located at the anterior part of the spinal canal or the presence of septae, which are uncommon in arachnoid cysts, may aid in the diagnosis of this type of cyst.3

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Regardless, the diagnosis of a neurenteric cyst can only be confirmed after histopathological examination.

Under the World Health Organization classification, a neurenteric cyst is described as composing mucin-secreting epithelium resembling that of the gastrointestinal tract. Based on the histological features of the cyst wall and its contents, according to the modified Wilkins and Odome classification, the cyst can be classified into three types. Type I cysts are the simplest, where the cyst wall consists of a basement membrane supporting a single, or pseudostratified, layer of cuboidal to columnar epithelial cells with or without cilia. Type II cysts have more complex glandular elements of the gastrointestinal tract or respiratory tree, including mucus glands, serous glands, fat, cartilage, bone, elastic fibers, lymphoid tissue or nerve ganglion, and smooth muscle in their wall. Type III cysts have ependymal or glial tissue in addition to the elements seen in the type II cysts. Most neurenteric cysts are type I, as in our patient.

Early surgical intervention is the only available effective treatment for intradural extramedullary lesions, and it is usually curative with good prognosis. Left untreated or with only partial cyst wall excision, the lesion may grow and cause severe arachnoiditis with irreversible damage to the spinal cord. Furthermore, the risk of rupture resulting in dense adhesion and scar tissue formation involving the dura further complicates the removal of any recurrences. The lesion may be fatal as a result of its dorsal displacement and compression of the brain stem; hence, aspiration of the cyst only is not recommended. Long-term follow-up is mandatory for incompletely excised cysts because they recur not infrequently. The osmotic effect of secretion from the partially excised cyst wall may contribute to the recurrence of the cyst with fluid accumulation.

Although the cyst is almost always located ventral to the spinal cord, laminectomy is the preferred approach during surgery. Other approaches such as transoral, transpalatal, suboccipital and pos-
terolateral transcondylar have been documented in the management of craniocecal and cervicothoracic junction neurenteric cysts. In conclusion, neurenteric cysts are uncommon tumours of developmental origin. Presentation ranges from being asymptomatic to profound neurological deficits, and is dependent on its location. Early surgical intervention and long-term follow-up in partial excision is recommend, particularly to prevent long-term morbidity.

Anterior C1-2 osteochondroma presenting with dysphagia and sleep apnea

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**Abstract**

Spinal osteochondromas are rare, benign tumors of the bone. These tumors can manifest as solitary lesions or as part of a hereditary syndrome. Most spinal osteochondromas occur in the posterior cervical spine and can cause myelopathy or radiculopathy. Osteochondromas of the anterior cervical spine that cause respiratory or swallowing symptoms are rare. We present the unique case of a solitary osteochondroma of the anterior C1 vertebral arch causing obstructive sleep apnea and dysphagia in a 16-year-old female. The patient underwent resection of the tumor via a left anterior transcervical approach to the spine. The patient’s symptoms resolved completely after surgery. To our knowledge, this is the first case of a sporadic osteochondroma arising from the anterior arch of the C1 vertebra causing dysphagia and obstructive apnea in a pediatric patient.

1. Introduction

Osteochondroma is a benign tumor of the bone that can be solitary or part of a hereditary syndrome known as multiple hereditary exostoses (MHE). Most spinal osteochondromas occur in the posterior elements of the cervical spine. Osteochondromas arising from the anterior elements are rare. In this report, we present the case of a sporadic osteochondroma arising from the anterior arch of the first cervical vertebra (C1) presenting with dysphagia, snoring, and sleep apnea in a 16-year-old girl without MHE.

2. Case report

Two years prior to presentation, the 16-year-old female noticed difficulty swallowing and the new onset of snoring. On physical examination, there was a visible bulge behind her pharynx. MRI and CT of the cervical spine revealed a bony lesion projecting anteriorly from the left side of the C1 arch and extending caudally to the second cervical vertebra (C2) body (Figs. 1 and 2). The lesion had high signal intensity on T1- and T2-weighted MRI and was consistent with osteochondroma. Over the next two years, her symptoms worsened and a repeat MRI revealed that the lesion did not grow but had lost its attachment to the C1 arch. The patient elected to have the lesion removed surgically because she felt the lesion was causing her symptoms. A transoral operation was initially considered because of the lesion’s location at C1; however, an en bloc resection would require a mandibular osteotomy. Given the benign nature of the lesion, a left anterior transcervical approach to the spine was chosen instead.

A transverse incision was used and cranial nerves XI, X, XII, the superior laryngeal neurovascular bundle, carotid artery, internal jugular vein, and great auricular nerve were identified and preserved. The tumor was identified in the retropharyngeal space (Fig. 3). A straight osteotome was used to make a cut at the base of the tumor and the bulk was removed in one piece. Further removal of tumor was performed in a piecemeal fashion with rongeurs. All gross tumor was removed except for the small stump emanating from the anterior arch of C1. Because of the benign nature of osteochondroma, we did not pursue this small stump due to the extreme angle of approach and risk of vascular injury. Histological analysis confirmed the diagnosis of osteochondroma. The patient did well postoperatively with minimal dysphagia that resolved after 2 weeks. A postoperative CT showed removal of most of the tumor except for the small remnant intentionally left at the arch of C1. At 10-month follow-up CT, there was no evidence of tumor growth, and the patient had significant alleviation of her symptoms.

3. Discussion

Osteochondroma is the most common benign bone tumor, and usually occurs in long bones. Osteochondromas are thought to arise from a lateral displacement of a portion of the epiphyseal growth cartilage to become a bone-like outgrowth capped by cartilage. Spinal osteochondromas are uncommon, and most cases occur in patients with MHE.