

# CASE REPORT

## TORTICOLLIS CAUSED BY NEUROENTERIC CYST OF UPPER CERVICAL REGION

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### Abstract:

#### Objective:

Torticollis is a symptom that can be related to different pathological mechanisms ranging from simple to life-threatening conditions. Here we report a child with torticollis caused by a neuroenteric cyst in the upper cervical region; this is a very rare condition in childhood and in this case, it was successfully resolved by surgery.

#### Clinical presentation:

A 2.5 year old boy presented with a 2 month-history of torticollis, he had developed paraparesia 2 weeks before admission. At examination he was found to be quadriparetic. Radiographic study of the cervical spine revealed widening of the cervical canal. Brain and spinal magnetic resonance imaging revealed a hypointense lesion on the T1 at the craniovertebral junction having a compressive effect on the anterior aspect of the brain stem and spinal cord.

#### Intervention:

The patient underwent surgery. After craniotomy and opening of the dura, a cystic lesion was seen; clear fluid was aspirated and the cyst wall was removed.

#### Conclusion:

Considering the quadriparesis and torticollis, the patient improved significantly within the first few days after surgery. No relapse of symptoms occurred during the follow up period. This is the first case report of a child in whom torticollis was due to a neuroenteric cyst of the upper cervical intradural region.

**Keywords:** Torticollis - Neuroenteric cyst - Child - Quadriparesis.

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### Introduction

Torticollis, an unusual position of the head and neck in tilt, rotation and flexion, is considered a sign rather than a specific diagnosis (1, 2). A wide spectrum of disorders, ranging from simple to life-threatening conditions, can provoke an acquired torticollis (3). Torticollis can be classified as being muscular or nonmuscular in origin (1, 2), and more than 80 different causes of torticollis have been described (4). Differential diagnosis may be difficult.

Neuroenteric cysts are rare congenital anomalies usually presenting as a posterior mediastinal mass. The occurrence of such lesions and compression of the spinal cord in the upper cervical region is a very rare phenomenon (2, 3, 4); it has other

names, such as teratomatous cyst and enterogenous cyst, probably reflecting uncertainty as to the etiology (5). Here we report a child with a neuroenteric cyst, who presented with recurrent episodes of torticollis and progressive quadriparesis. The condition was resolved successfully with total removal of the cyst.

**Case report**

The patient was the 2.5 year-son of noncosanguineous, healthy parents, without any family history of trauma. He was delivered by normal vaginal delivery and had no history of perinatal complications. The boy was well until 2 months prior to admission, when he presented with an episode of torticollis lasting one week and weakness of extremities particularly the right upper limb; these symptoms became progressively worse, accompanied by ataxia. Initially torticollis had disappeared for 10 days, but then became persistent until admission. Torticollis did not worsen under conditions of stress and

the patient never reported muscular pain. On examination the boy looked well. The right upper extremity was paretic (muscle power 1/5); the left hand and both feet were paretic (3/5). Deep tendon reflexes were brisk bilaterally. There was a vague sensory impairment of all modalities of sensations with a loss of pinprick sensation to the level of the upper cervical region. Neither spinal tenderness nor sternocleidomastoid contraction was noted. Babinski's sign and ankle clonus were detected. He was unable to sit, stand, or walk because of a severe spastic quadriparesis. X-rays showed widening of cervical canal and pedicle erosion (fig 1) and cervical MRI showed a hypointense lesion on T1, at the craniovertebral junction, having compressive effects on the anterior aspect of brain stem and spinal cord (fig 2). Following laminectomy and craniotomy, a cystic lesion protruded through and after aspiration, was removed completely (Fig 3).

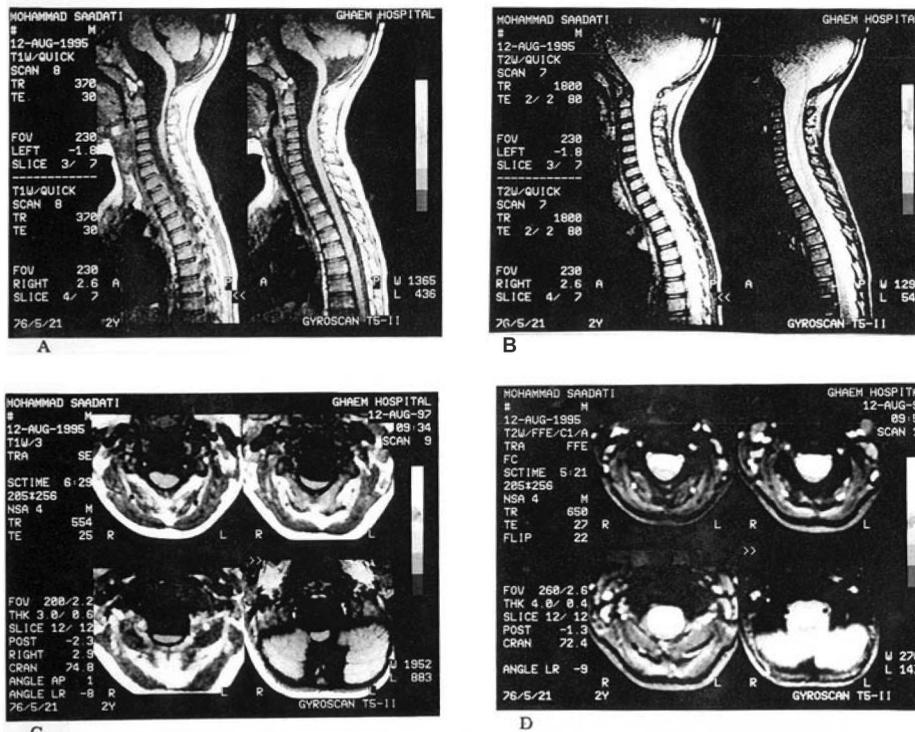


Figure 2  
Neurenteric Cyst typically situated ventral to medulla and spinal cord junction appears hypointense on T1 (A) and hyperintense on the T2(B),C . Axial T1, D . Axial T2 weighted MRI

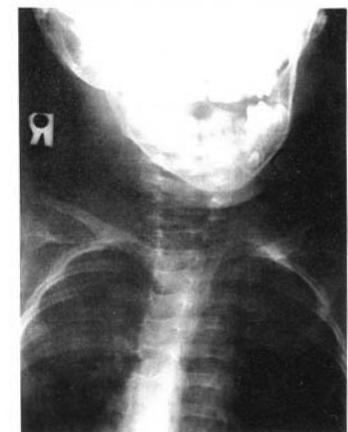


Figure 1  
Frontal view of cervical spine demonstrates widening of the interpediculate distance in upper cervical spine.

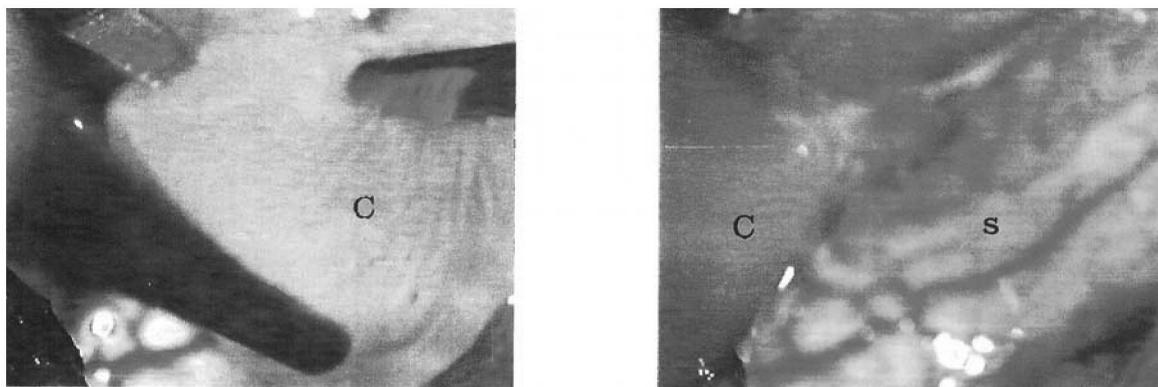


Figure 3: Intraoperative microscopic views of neuroenteric cyst  
C . cyst , S . spinal cord



Post operatively motor power improved significantly on the first day. The patient was discharged on the 10th day with fair motor function and no torticollis and now walks with little support. The pathological finding was a cystic lesion about 3.5 X 1.5 centimeters, with the cyst wall being composed of an outer coat of collagen supporting basement membrane and an inner layer of cuboid epithelium containing positive periodic acid-schiff mucin.

### Discussion

Neuroenteric cysts are rare malformations that may lead to spinal cord compression or tethering (6). The name enterogenous cyst was first applied by Harrimon to intraspinal, usually extramedullary cysts, lined by columnar epithelium of intestinal character (7). Embryogenesis of enterogenous cysts is a matter of debate. Rhaney and Barclay suggested that the intra spinal enterogenous cyst was a product of abnormal separation of germ layers. 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. In the last stage,

when separation (excalation) occurs, groups of endoderm cells may be carried back with the mesoderm, giving rise to enterogenous cysts (8). The neuroenteric cyst may present at any age. There is a 3:2 male predominance. Our patient was a 2.5 year-old boy.

The complex association of intestinal protrusion, cloacal or bladder extrophy and renal dysgenesis may be fatal. The presentation of intra spinal cysts is more common during adulthood. The symptoms of spinal cord or nerve root compression may mimic those of the space occupying lesion (9, 10).

Our patient demonstrated a peculiar clinical course, characterized by tilting of the neck to the left side. During 2 months, the patient underwent several sessions of physiokinesis treatment without success. He was referred to us when further neurological disturbances appeared, including quadriparesis and ataxia. Imaging detected a compressive cystic lesion on the spinal cord and brain stem. This case clearly confirms the need to consider central nervous system abnormalities in any child with recurrent or unexplained torticollis, because it may represent a life-threatening condition that demands

prompt evaluation and treatment(9). Torticollis maybe related to several diseases but is usually a simple condition in children; however when it is accompanied with other neurologic signs or symptoms, life threatening conditions should be considered (10). In view of the clinical findings, more than one cause, such as compression of the right spinal accessory nerve and mass effects on brain stem may be considered in our patient; in this case, abnormal clinical findings disappeared a few days after surgery. Several authors claim that the rapid clinical improvement is related to the removal of the mass effect exerted by cyst.

In conclusion, this is the first description of a child with a peculiar recurrent torticollis, and subsequent progressive paraparesis. The clinical history of this patient should alert pediatricians to consider a possible neurological disorder in any child with unexplained torticollis.

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