

# Acute Tetraplegia after Trivial Neck Injury Revealed a Cervical Intraspinal Neurenteric Cyst in a Child

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## Abstract

Neurenteric cysts (NC) are rare anomalies of the central nervous system (CNS). They result from abnormal separation of the neural tube and the endodermis during the 3rd week of gestation, which leads to the persistence of endodermal elements in the spinal cord. They usually present insidiously with local spinal pain, radiculopathy, and myelopathy. We report the case of an 8-year-old boy with a high cervical neurenteric cyst who developed a progressive myelopathy after a minor neck trauma. He recovered completely after cyst removal.

**Keywords:** Neurenteric cyst; Injury; Spinal cord tumor

## Introduction

NC is rare anomalies of the CNS. They result from abnormal separation of the neural tube and endodermis during the 3rd week of gestation, which leads 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].

NC represents 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, comprising symptoms and signs of progressive myelopathy [6]. Decompressive surgery is the treatment of choice. The outcome is usually good because of the mild preoperative symptoms. We report a case of intraspinal cervical NC that presented as acute tetraplegia after trivial neck injury. A full neurological recovery was achieved with surgical resection of the NC.

## Case Presentation

The patient and his parents were informed that information concerning the case would be submitted for publication.

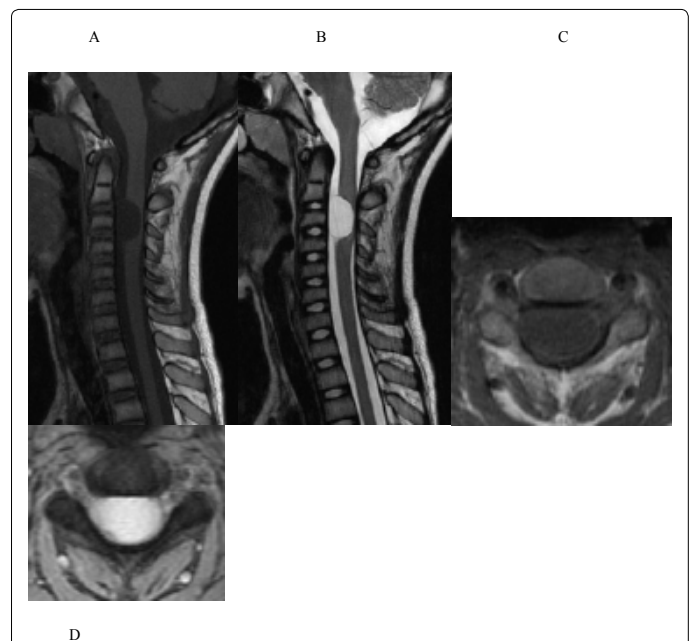
An 8-year-old boy presented with a delayed myelopathy after a minor neck injury. Ten days prior to referral to our department, his mother's elbow accidentally struck the back of his neck. There was only mild posterior neck discomfort on the night of the injury. Over the next 4 days, a slowly worsening tetraparesis developed, associated with some increase in neck pain. He was admitted to the nearest hospital and evaluated with spinal Magnetic Resonance Imaging (MRI) and referred to our department with the diagnosis of a spinal tumor. He showed rapidly progressive gait disturbances and urinary hesitancy, characterized by decreased flow and cessation of urination in mid-stream.

He had an unremarkable medical history and no significant history of trauma to the spine, except the mild strike to his neck with his mother's elbow. The family history was negative for congenital spinal or neurological conditions.

His posterior neck was slightly tender to palpation in the high cervical area, but demonstrated no swelling. Motor dysfunction rapidly progressed after emergency hospitalization. He showed tetraplegia and urinary retention a few hours after the hospitalization to our department. Sensory findings revealed complete loss without vibration sensation in his feet.

## Imaging

Plain cervical spine films, including lateral, anteroposterior, and odontoid views, failed to reveal any abnormality. The emergency MRI study showed a large intradural extramedullary cystic lesion in the spinal canal extending from C2-4, with its greatest dimensions adjacent to the C3 vertebral body (Figure 1). The lesion was ventral and lateral



**Figure 1:** MRI study. T1 weighted images (a,c). T1 weighted images (b,d). MRI showed a large intradural extramedullary cystic lesion in the spinal canal extending from C2-4. The lesion was ventral and lateral to the right in relation to the spinal cord, causing marked compression and displacement of the cord, especially adjacent to C3. The lesion was of a similar intensity to CSF on both T1 and T2 weighted images.

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to the right of the spinal cord, and was causing marked compression and displacement of the cord, especially adjacent to C3 (Figure 1). The lesion was of a similar intensity to cerebrospinal fluid (CSF) on both T1-weighted (spin echo 600/10) and T2-weighted (spin echo 2200/90) imaging sequences (Figure 1). It failed to demonstrate enhancement with intravenous gadolinium-DTPA administration.

### Operation

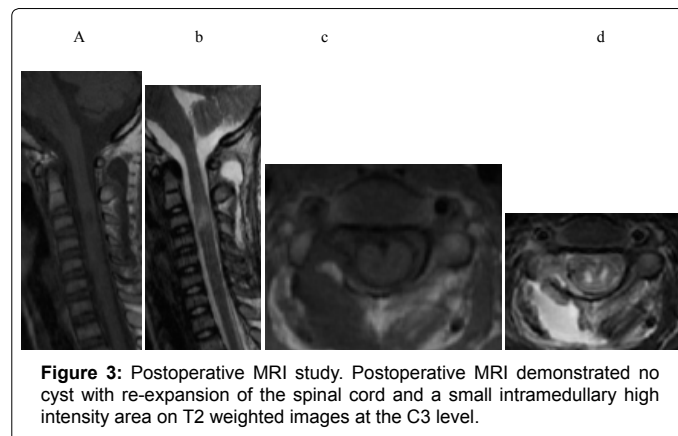
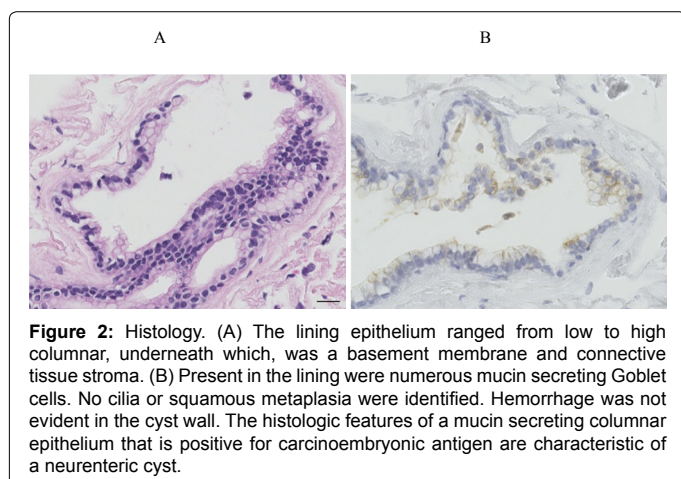
The patient was taken to the operation room upon completion of the MRI. After careful bronchoscopic intubation and Mayfield pin head fixation he was turned prone. A cervical hemilaminectomy from C2-4 and medial facetectomy from C3-4 were performed. The paraspinous muscles and ligaments appeared non-traumatized. The right head of the C2 spinous process was cut and preserved. It was put back and fixed with a suture after intradural maneuver to prevent postoperative cervical instability. A taut cyst was revealed after dural opening. It was punctured and aspirated with a syringe, yielding about 3 mL of clear CSF-like fluid without any trace of blood. This immediately slackened the cyst. The cyst was ventrolateral on the right, displacing the spinal cord dorsal and lateral to the left. The cyst wall was thin and translucent. This lining was not septated and quite adherent to the ventral surface of the cord in the middle. An excision of the cyst was performed, removing the great majority of the lining, except for adherent material on the ventral surface of the cord.

### Histology

Microscopically, the lining epithelium ranged from low to high columnar, beneath which, was a basement membrane and connective tissue stroma (Figure 2a). Present in the lining were numerous mucin-secreting Goblet cells. No cilia or squamous metaplasia were identified. Hemorrhage was not evident in the cyst wall. The histologic features of a mucin-secreting columnar epithelium that is positive for carcinoembryonic antigen are characteristic of a neurenteric cyst (Figure 2a and 2b).

### Post-operative and follow-up

The patient experienced an uneventful post-operative course. He showed progressive recovery of tetraparesis, and he could ambulate on the seventh post-operative day. Urinary symptoms disappeared post-operatively. Two weeks after surgery neurological examination showed no detectable weakness and only subtle Romberg's sign. Post-operative MRI demonstrated no cyst with re-expansion of the spinal cord and a small intramedullary high intensity area on T2 weighted images at the C3 level (Figure 3). He was clinically normal 3 months after the surgery.



### Discussion

NCs are rare congenital cysts of the spinal canal and are lined with epithelium of endodermal origin. Their exact embryopathogenesis is not well understood, but they result from a disruption occurring in the 3rd week of embryogenesis and involve the complex series of interactions responsible for the formation of the notochord, neuroenteric canal, endoderm, and neural tube [2,7].

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 lesions [8]. They can be extradural or intradural, and occasionally intramedullary [9]. They are frequently associated with several vertebral anomalies: hemivertebra, absence of the vertebra, anterior and posterior spina bifida, and kyphosis [4,10]. They can also be associated with diastematomyelia or syringomyelia [4,10]. Intrathoracic or intra-abdominal cysts can coexist with NCs with a fibrous band joining them to the intraspinal NC through a vertebral defect [11]. An open communication between the two cysts is rare.

In the case presented here, the NC was in an atypical site at the high cervical region, but at a usual location anterior to the spinal cord without vertebral fusion anomaly.

The clinical spectrum of NC depends on the site of the lesion and includes local pain, radiculopathy, and/or myelopathy [3,6]. Neonates and young children frequently present with symptoms related to an intrathoracic or intra-abdominal cyst; they rarely lead to imaging findings suggestive of meningitis [12]. 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 [13]. Progressive clinical manifestations may follow trauma [14]. The possible explanations for these unusual presentations 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 [3,14].

In the present case the onset of symptoms was related to a minor neck trauma. Worsening pain and a progressive myelopathy developed over 10 days. Of significance is that the patient was neurologically intact immediately after and for approximately 24 h following the trauma. The cyst contained clear fluid without any discernible hemorrhage. There was no visible bruising of the cyst wall or spinal cord, and on pathological examination of the cyst wall, no hemorrhage was found.

On analysis, this information excludes hemorrhage into the cyst as a possible event in the pathogenesis of the deterioration. It also casts doubt on an acute mechanical compression at the time of injury as being the pathophysiologic mechanism. Instead, the delayed and progressive neurological deterioration suggests a more dynamic process. Progressive cyst expansion and cord compression related to altered fluid dynamics, with accumulation of CSF and/or cyst fluid. Consequently, the progressive ischemia, and therefore dysfunction, of an already compromised spinal cord by the compression of the NC finally reached a threshold level that impaired the local microcirculation.

In summary, intraspinal congenital cysts may occasionally present with a rapid onset and quick progression of myelopathy after a minor spine injury. We speculate that a small increase in the cyst size secondary to the trauma results in increased mechanical distortion and spinal cord dysfunction on a compressive and ischemic basis.

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