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CASE REPORT

# Rare morbidity of permanent quadriplegia caused by neurenteric cyst of the cervical cord

Henry Tan Chor Lip, Tan Jih Huei, Ariz Chong Abdullah, Noor Azman Bin A. Rahman

*Department of Neurosurgery, Hospital Sultanah Aminah Johor Bahru, Johor, Malaysia*

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## ABSTRACT

The incidence of neurenteric cyst (NC) is rare, accounting for 0.3%–1.3% of all spine tumors. The occurrence of quadriplegia caused by NC is even scarcer. Herein we report on a young girl with a rare NC over the C2-C5 spinal cord, which led to the morbidity of permanent quadriplegia despite early surgical intervention. This case highlights the rare morbidity of cervical cord NC presenting with permanent quadriplegia that failed to respond despite early surgical excision.

**Key words:** Cervical cord, laminectomy, neurenteric cyst, quadriplegia

## INTRODUCTION

Neurenteric cyst (NC) is a rare spine tumor commonly located in the cervicothoracic segments of the spinal cord.[1] It is a congenital anomaly caused because of the persistence of the neurenteric canal and commonly detected only in the second and third decade of life.[2] Surgical outcomes are often curative with only 11% experiencing worsening of symptoms because of cyst recurrence.[3] From our literature search, only one case was reported of acute-onset quadriplegia because of neurenteric cyst.[4] Herein we report on a similar case of 16-year-old girl diagnosed with NC over the C2-C5 segments of spinal cord with quadriplegia and on postsurgical excision via posterior approach remained in a permanent quadriplegic state, requiring home ventilator support. This case highlights the uncommon case of NC with its rare morbidity of permanent quadriplegia despite early surgical intervention.

## CASE PRESENTATION

A 16-year-old girl presented with initial symptoms of posterior midline neck pain for 6 days to the neurosurgery department. She was initially treated with analgesics by the primary care physician for muscle spasm. Throughout the span of 6 days prior, progressive quadriplegia with worsening respiratory distress was observed. No histories of trauma, upper respiratory tract infection (URTI), diarrhea, fever, meningism or raised intracranial pressure were observed before the onset of symptoms. The patient was intubated due to worsening respiratory distress and nursed in the intensive care unit. Physical examination revealed loss of power (Medical Research Council grade, 0/5 over upper and lower limbs) and absent sensation from C4 dermatome and below. Deep tendon reflexes were normal over the upper limbs (biceps, brachioradialis, and triceps) and hyperreflexia over the lower limbs (patellar and Achilles tendon). Cranial nerves were preserved and intact. Hoffman sign and Babinski sign were negative. Systemic examination did not reveal any abnormalities, and vital monitoring was within normal limits. The white cell counts were within normal limits, $6.9 \times 10^9$/L (4.0–11.0). The full blood count, blood urea, electrolytes, and creatinine were all within normal limits. An urgent magnetic resonance imaging (MRI) of the brain and spine revealed an intradural extramedullary mass anterior to the cervical spinal cord from the level of lower border of C2 to the upper

**Address for correspondence:** Dr. Henry Tan Chor Lip, Department of Neurosurgery, Hospital Sultanah Aminah Johor Bahru, Jalan Persiaran Abu Bakar Sultan, 80100, Johor Bahru, Johor, Malaysia.
E-mail: relos1402@gmail.com



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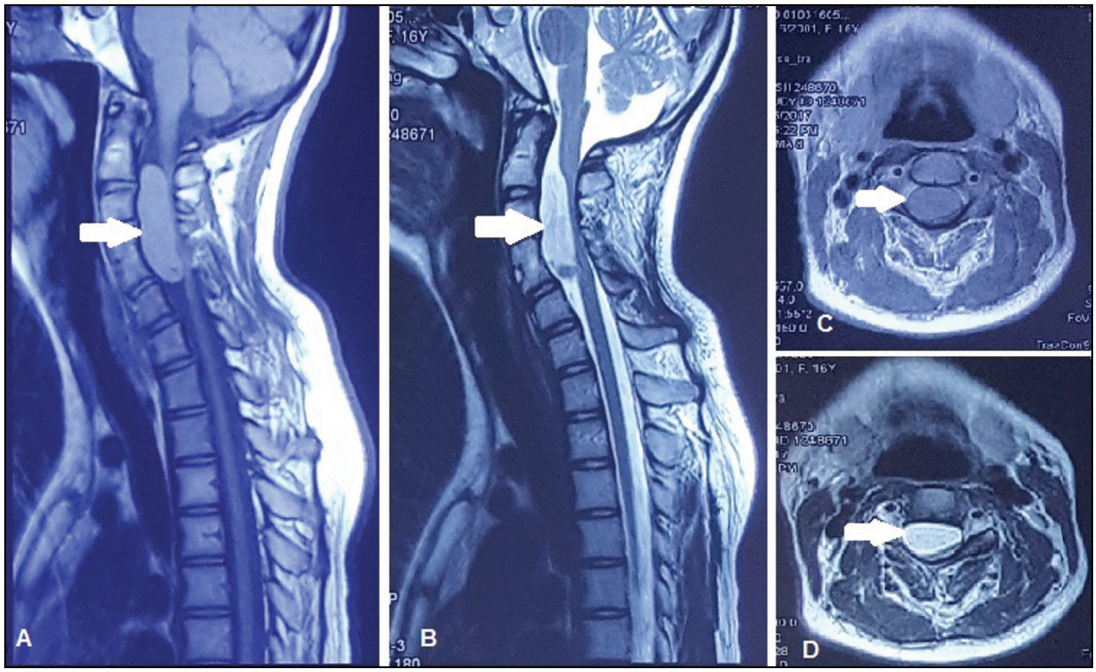


**Figure 1:** Magnetic resonance imaging of spine showing neurenteric cyst (white arrow) from lower border of C2 to upper border of C5, which is isointense on T1-weighted and hyperintense on T2-weighted images. (A) Sagittal T1. (B) Sagittal T2. (C) Axial T1. (D) Axial T2. The neurenteric cyst is causing severe compression and narrowing to the spinal cord at C2-C5 segments.

border of C5, causing compression to the spinal cord. The mass was isointense on T1-weighted and hyperintense on T2-weighted images [Figure 1]. No focal lesions were observed within the brain parenchyma. On the same day of admission, an urgent laminectomy was performed from the level of C2-C5. The dura was opened from C2-C5, and the spinal cord was visualized with an intraoperative microscope. The cord appeared tense and bulging in the midline with absent pulsations. With gentle retraction of the arachnoid layer, a cyst was identified as the cause of compression to the spinal cord anteriorly. The content of the cyst was aspirated first to reduce its size and the tension on the spinal cord. Approximately 4 $cm^3$ of a thick, non-foul smelling, yellowish mucoid fluid was aspirated using a 16G branulla [Figure 2]. No presence of hemorrhage or signs of infection was observed within the cyst on aspiration. With subsequent meticulous dissection, the cyst was successfully dissected away and excised entirely from the spinal cord. After excision, the cord appeared relaxed and pulsatile. Gross examination revealed a yellowish well-encapsulated cyst measuring $4 \times 1.5 \times 1$ cm. Histological examination revealed that the cyst wall was composed of simple cuboidal to columnar mucinous epithelium [Figure 3]. No cilia and goblet cells were identified. The histology features of a mucin-secreting columnar epithelium were consistent of an NC. Furthermore, the fluid aspirated from the cyst cultures was sterile and tested negative for *Mycobacterium tuberculosis*. Postoperatively, the patient remained in a permanent quadriplegic state without any improvement in power or sensation. Despite performing a tracheotomy, she remained ventilator dependent, most likely because of the permanent damage to the C3, C4, and C5 spinal cord, which paralyzed the diaphragm. Prior discharge, she underwent rigorous physiotherapy and rehabilitation exercises to prevent early contractures. She was discharged with a portable home ventilator after her caretakers were self-reliant on taking care of the patient with a total length of stay of 2 months. Physiotherapy exercises were also taught to the patient's caretakers so that they may continue doing it at home.

## DISCUSSION

Spinal NCs are rare and comprise 0.3%–1.3% of all spine tumors.[1] The formation of NC is due to a congenital anomaly from the persistence of neurenteric canal that prevents the separation of endoderm and notochord. Remnants of neurenteric canal lead to the formation of NC, which is defined by the presence of mucus-secreting epithelium, resembling cells of the gastrointestinal tract.[2] Majority of NC occurs within the cervicothoracic segments ventrally to the spinal cord. Uncommon locations of NC have been reported at the craniovertebral junction, cerebellopontine angle, and supratentorial space.[5] Patients commonly exhibit symptoms of progressive local tenderness at the level of spinal axis pathology, myelopathic symptoms, and muscle weakness at second and third decades of life. Male patients are more predisposed to NC with a male-to-female ratio of 2:1.[2] MRI remains the gold standard for diagnostic imaging for NC. Despite a variable number

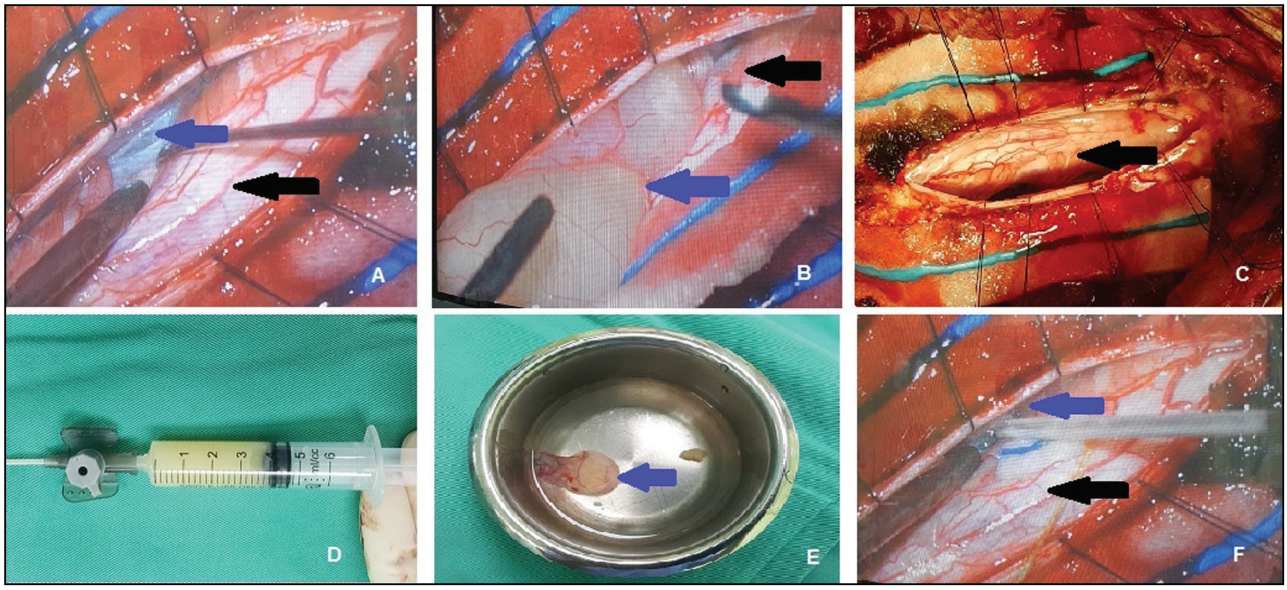


**Figure 2:** (A) Intraoperatively, the cyst was identified (blue arrow) with gentle manipulation of the tensed spinal cord (black arrow). (B) The well- encapsulated cyst (blue arrow) was excised entirely exposing the tensed spinal cord (black arrow). (C) Spinal cord (black arrow) was loose and relaxed after excision of cyst. (D) Yellowish fluid aspirated before excision. (E) Neurenteric cyst (blue arrow) completely excised. (F) Neurenteric cyst (blue arrow) was aspirated using 18G branulla exposing the loose spinal cord (black arrow).

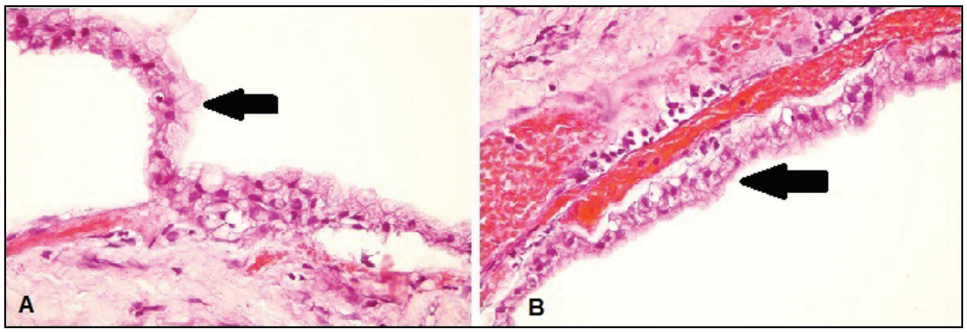


**Figure 3:** (A) and (B) Histological section of the cyst wall composed of simple cuboidal to columnar mucinous epithelium (black arrow) compatible with a neurenteric cyst

of MRI reports in the literatures, the majority of NC is characterized by a non-contrast-enhancing mass that is isointense on T1-weighted sequences and hyperintense on T2-weighted sequences, which was similar to the findings observed in this patient.[2,6] The first-line treatment is for complete surgical excision to decompress the pressure over the spinal cord.[7] Complete excision is important to prevent cyst recurrence from a partial resection. There are three surgical techniques (anterior, posterior, and lateral) to approach the spine lesion, with the posterior approach being widely practiced. The posterior approach is adopted due to its fewer intraoperative complication risks.[3] Most complications arise because of the process of laminectomy, which may cause injury to the dura, spinal cord, and nerve roots with the formation of hematoma due to epidural venous bleeding. The option of cyst aspiration before excision should be carefully considered intraoperatively as it can cause meningeal irritation and meningitis because of leak of cyst contents postoperatively.[8] However, aspiration may be performed to reduce the cyst size and its potential risk at further cord damage during excision by relieving the tension placed on the spinal cord. In our case, the cyst was aspirated first because of its great pressure on the spinal cord and to reduce potential further damage to the spinal cord during excision. Surgical outcomes are generally curative of the sensory and motor deficits with only 11% of cases experiencing worsening of symptoms pre-morbidly in cases of myelopathic signs that were treated early.[3] From our literature search, this is the second case being reported of NC causing permanent quadriplegia, which represent significant morbidity in a young adolescent patient. The possible causes of acute symptoms considered were possibility due to Guillain–Barré syndrome and spinal cord compression.[9] In our case, the cause of spinal cord compression was due to the NC. It has been reported that hemorrhage or infection could cause acute spinal cord compression. However in our case, we postulate that the symptoms were due to the mass effect of the NC itself on the spinal cord. This is evidenced by the absence of hemorrhage and negative culture yield of the cyst content.

This patient had no symptoms of infection (URTI/acute gastroenteritis) 2 months before symptoms. Her clinical symptoms correlated with the level of spinal cord compression, which was identified on the spine MRI. Intraoperatively, the spinal cord had evidence of compression whereby the cord appeared tense and bulging without any pulsations, which indicated significant compression. The presentation may mimic Guillain–Barré syndrome, however in the acute setting, without the presence of infection and gross evidence on the spinal cord imaging, the most likely diagnosis was due to a spinal cord compression because of NC.[9] This case is important to highlight the rare morbidity of NC over the cervical region, which may cause permanent quadriplegia despite early surgical treatment.

## CONCLUSION

From our literature search, NC over the cervical cord may represent with trivial neck pain, and surgical outcomes are often optimistic for most patients. However, our case is an example of rare outcome of permanent quadriplegia, which occurred in a very young adolescent female with great disability. Counseling is necessary to include permanent quadriplegia as the possible complications during preoperative consent taking.

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### Conflicts of interest

There are no conflicts of interest.

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