

CASE REPORT

A Rare Case of Neurenteric Cyst of Spinal Cord with Thoracic Vertebra Fusion Successfully Managed with Spinal Cyst Excision and Posterior Instrumentation Surgery

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ABSTRACT

The incidence of neurenteric cyst (NC) is rare amongst spine tumors. It is most often asymptomatic but may present with sensory and motor symptoms. When associated with thoracic vertebra fusion it is not reported before, this complicates the placement of pedicle screw during posterior instrumentation. Herein, we report a case of thoracic spinal neurenteric cyst in a 40-year-old man that presents with chronic back pain, left lower limb weakness and numbness. Elective excision of NC over T6-T7 with laminectomy and multilevel posterior instrumentation was successfully performed with significant improvement of the symptoms. Neurenteric cyst is a rare spinal cord lesion which may cause permanent neurological sequelae. Complete surgical excision with spine fixation in this case provides good long-term outcome.

Keywords: Neurenteric cyst, Thoracic spine, Vertebra fusion, Spine surgery

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INTRODUCTION

Neurenteric cyst (NC) is a rare intradural extramedullary spine tumor. It is commonly found within the cervicothoracic region of the spinal cord. (1) It is hypothesized to be caused by a congenital malformation due to 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 due to cyst recurrence. (3) This report describes the clinical course and management of the one rare case of NC with associated thoracic vertebra fusion.

CASE REPORT

A 40-year-old male presented to the outpatient neurosurgery clinic for progressive back pain for one month which radiates to both lower limbs. The pain was aggravated with coughing and resulted in insomnia. There was an associated bilateral lower limbs weakness, notably more so over the left side with muscle power of 4/5. Neurologic examination revealed both lower limbs reflexes were brisk with an increased in muscle tone. Sensations were reduced from the T8 dermatome

and below. Thoracic spine radiograph revealed a fused T7 to T9 vertebra bodies. Magnetic resonance imaging T1-weighted images (T1WI) demonstrated a T6 to T7 lesion displaying intermediate signal intensity without enhancement after gadolinium administration. The lesion was hyperintense on T2-weighted images (T2WI) (Fig. 1). Laminectomy of the T6 to T7 vertebrae was performed, which revealed a circumscribed, semitransparent pale cyst that was located ventrally to the spinal cord. The cyst was completely excised from its arachnoid attachment. Aspiration of the cyst yielded a yellowish mucoid liquid. (Fig. 2) The fluid was sterile and tested negative for acid fast bacilli. Posterior instrumentation with pedicle screws was performed at T4, T5, T8, T10 level to stabilize the spine. (Fig. 3) Histopathologic examination of the surgical specimen was consistent with the features of a neurenteric cyst. The cystic lesion was lined by pseudostratified, ciliated columnar epithelium with occasional goblet cell seen. (Fig. 2) After surgery, both the back and leg pain were both ameliorated. Follow up at 1 year, he remains symptoms free.

DISCUSSION

Spinal neurenteric cysts (NC) are rare and comprises of 0.3%-1.3% of all spine tumors (1). The formation of NC is due to the persistence of neurenteric canal that prevents the separation of endoderm and notocord (2). Majority of NC occurs within the cervicothoracic

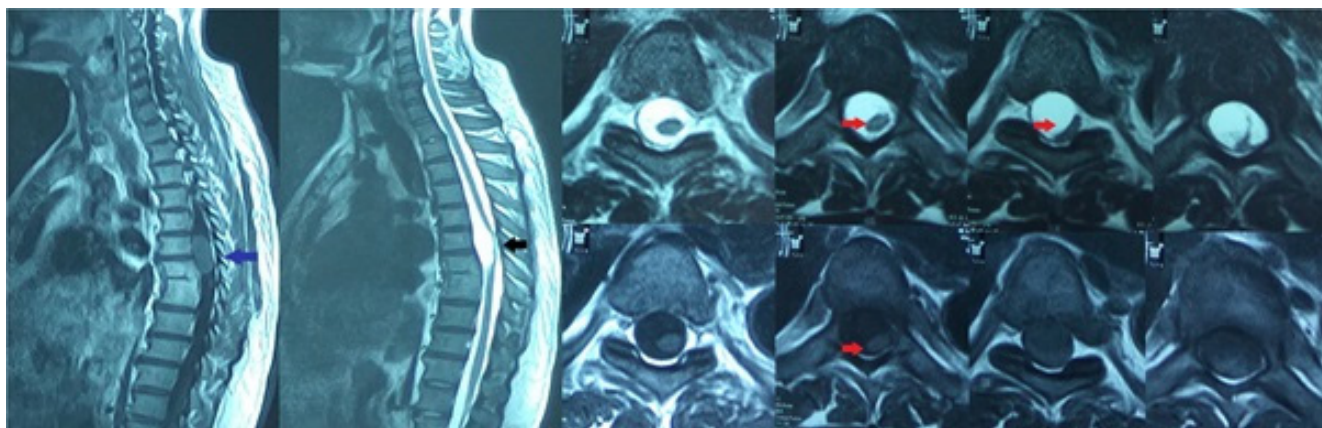


Figure 1: MRI of spine showing neurenteric cyst from T6 to T7 which is isointense on T1 (blue arrow) and hyperintense on T2-weighted (black arrow). On axial view, the neurenteric cyst is causing compression and narrowing to the spinal cord (red arrow).

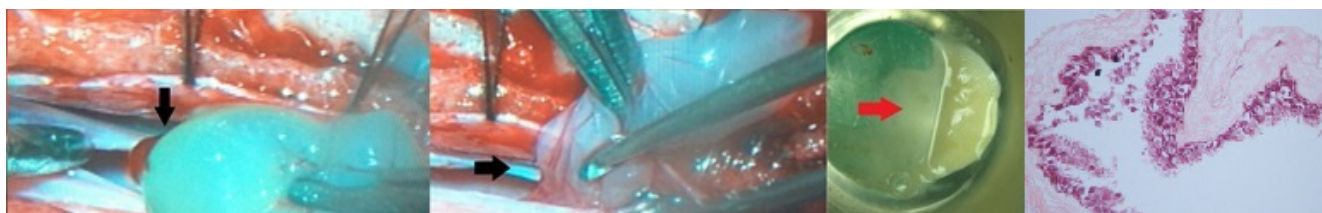


Figure 2: Intra-operatively the cyst was identified with its attachment to the adjacent arachnoid (black arrow). Yellowish fluid aspirated prior to excision (red arrow). Histological Slide of the resected cyst showing pseudostratified ciliated columnar epithelium with occasional goblet cell.

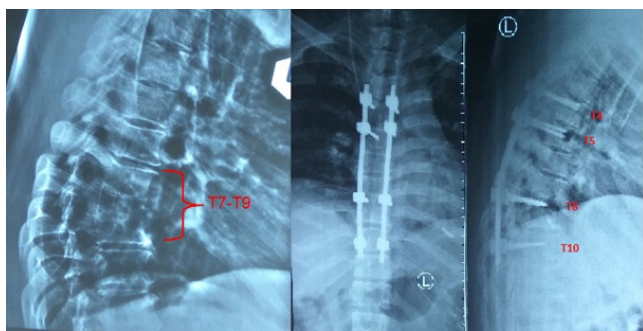


Figure 3: Thoracic Spine Radiograph preoperatively and postoperatively after posterior instrumentation at T4, T5, T8 and T10

segments ventrally to the spinal cord which is also seen in this case and other case series shown in table 1. We summarize all published series of neurenteric cyst of spinal cord in the tabulated form with regards to the year of report, number of patient and their characteristics, common location and recurrences rate with total or subtotal excision (Table I). From these previous reports, NC is more commonly seen in children and complete excision resulted in the best outcome with lowest recurrence. However, subtotal resection may be an alternative in critical area as the cyst run a benign clinical course (3).

Patients commonly present with symptoms of progressive local tenderness at the level of spinal axis pathology, myelopathic symptoms and muscle weakness at second and third decades of life (2). Surgical outcomes are generally curative of the sensory and motor deficits with only 11% of cases experiencing worsening of symptoms pre-morbidly (3). The extreme case when treated early may reverse the neurology. A case from Japan reported

Table I: List of published series of neurenteric cyst of spinal cord

Author and year of publication	Number of patients and characteristics	Most common location	Procedures performed and outcome
de Oliveira RS et al, 2005	16 children, aged 20 days to 14 years,	Ventral aspect of spinal canal (n=7). Spinal deformities in 5.	12 total resection- no recurrence 4 partial resection with 3 recurrence
Kim CY et al, 1999	8 cases (4 children)	7 in spinal cord, 1 at posterior cranial fossa	Total removal – no recurrence. Subtotal excision- no recurrence (follow up 38 months)
Chavda SV et al, 1985	8 patients (8 – 44 years)	lower cervical and upper thoracic spinal cord	37% risk of recurrence. Exact operation not mentioned.
Kimura H et al, 2006	18 cases of recurrent NC	CPA and cervical	16 cases following partial cyst resection 2 cases following total resection 37.1% (29/78) – total resection =>no recurrence 55.1% (43/78) – partial resection => 5 recurrence (11%) Other procedure- aspiration+ shunt / fenestration shunt
Paleologos et al., 2000	80 cases	cervical	Other procedure- aspiration+ shunt / fenestration shunt
Garg N et al, 2008	23 patients	Cervicothoracic Intradural extramedullary (n=16)	Partial excision – 63% recurrence (5/8)

a child presented with acute quadriplegia which did not progress to permanent quadriplegia (4). A neurenteric cyst is a part of the split cord syndrome. More than half of them can present with associated malformations such as vertebral deformities (particularly anterior spina bifida, hemivertebrae, spinal dysraphism, scoliosis, and Klippel-Feil syndrome), visceral or cardiac anomalies, anal atresia, syringomyelocele, meningocele and myelomeningocele. Clinical presentation of patients harbouring a solitary neurenteric cyst is mainly due to medullary compression. A neurenteric cyst is always attached to the spinal cord with a fibrous band. Therefore, its last portion may be adherent and may cause cord injury and traction unless divided.

MRI is the gold standard choice of imaging for NC. Despite a variable number of MRI reports in the literatures, the majority of NC is characterized by a non-contrast-enhancing mass that are isointense on T1-weighted sequences and hyperintense on T2-weighted sequences. The cyst mass is typically intradural and extramedullary with only 5% found intramedullary (2). This patient had typical radiological features of NC on the MRI images as reported in literature.

The first-line treatment for NC is complete surgical excision to decompress the pressure over the spinal cord. Complete excision is important to prevent cyst recurrence from a partial resection as shown in Table I. There are three surgical techniques (anterior, posterior and lateral) to approach the spine lesion, with the posterior approach being widely practiced despite the cyst typically being located anteriorly in relation to the spinal cord. The posterior approach is more widely practiced due to its fewer intra-operative complication risks. The anterior approach adds additional surgical complexity and an increased need for instrumented fusion, higher risk of damage to adjacent neurovascular anatomy, fusion failure, hematoma formation, and CSF leakage (2). In current case, we were able to gain sufficient access to the cyst from posterior approach, after minimal cord manipulation and cyst aspiration (3).

Interestingly, this case has a peculiar finding of fusion of the T7 to T9 vertebra body. This finding was not reported before. The vertebra changes reported were

pedicle or body erosion. This changes also is rare and seen in less than 5% of cases (5). The presence of spinal fusion have prompted the posterior instrumentation following the T6-7 laminectomy. As the T7 to T9 is fused, laminectomy of T6/T7 is predicted to have effect on T8 and T9 also with potential instability due to more than 3 vertebral level involvement [4 level = T6 + T7 (T7+T8+T9)]. Hence, the screw was fixed at pedicle of T4, T5, T8 and T10 vertebra stabilize the spine.(2) This case is the first to be reported with an association of idiopathic thoracic vertebra fusion.

CONCLUSION

Neurenteric cyst may present with incidentally or with mild local spine symptoms to acute and rapid deterioration of neurologic function. Total excision of spinal neurenteric cyst is essential to prevent recurrence. Vertebra fusion in the presence of neurenteric cyst is an unusual finding. Posterior instrumentation for spine stabilization may be considered when the total laminectomy segment more than 3.

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