Introduction:
The intraspinalenterogenous cyst, also called aneurenterogenous cyst, is a rare congenital disease. It was reported to be local to the C1 to L2 spinal segments, with the majority located in the cervicothoracic region. These cysts result from inappropriate segmentation of the notochord during embryogenesis causing endodermal tissue to remain in the spinal canal. It accounts for only 0.7-1.3% of spinal axis tumors. Only 12.2% of neuroenteric cysts are documented to be intramedullary.

Abstract:
Background: Neuroenteric cysts are rare non-neoplastic lesions arising from a failure of dissolution of the transient neuroenteric canal between the foregut and the notochord. They are most frequently seen in the intradurextramedullary space in the lower cervical and upper thoracic spine.

Case description: A 5 yrs old boy presented to us with the complaints of neck and upper back pain and weakness of all four limbs. MRI scan shows an intradurextramedullary cystic lesion at C6-T1 with significant compression over cord. After patient’s preparation a Posterior approach was used to remove the cysts. Post-operative course was un-eventful. Histological results were consistent with neuroenteric cysts. MRI image of 3 months follow-up shows no residual cysts and the boy has no further complaints.

Conclusion: Neuroenteric cyst (NC) is a rare lesion usual location at lower cervical and upper dorsal area and should be considered among differential diagnoses. Complete excision is the treatment of choice. In most instances a dorsal surgical approach will be satisfactory.

Keywords: Neuroenteric cyst, Laminoplasty, Cervicothoracic, Spinal cord tumor.

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Matson coined the term neurenteric cyst in 1954\textsuperscript{10}. The disease is officially named as enterogenous cyst in 1958 by Harriman\textsuperscript{11}. Previous studies of neurenteric cysts indicates that the disease is slowly progressive and rarely shows symptoms of acute onset\textsuperscript{1}. We report a case of cervical intradural extramedullary enterogenous cyst with severe clinical presentation.

**Case Report:**

A 5 yrs boy presented with gradually progressive neck and upper back pain with quadripareisis for last 1 yr. the pain was severe in intensity. On examination the baby was conscious and oriented; muscle power of upper limbs was normal excepts weakness in grip in both hand and lower limbs was 4/5. Diminished sensory level found at D4. All jerks were exaggerated. Hoffman’s sign was negative. Autonomic functions were normal.

MRI of cervico-dorsal spine shows well circumscribed elliptical in shape intradural cystic lesion extending from C6 to D2 level with significant cord compression. The lesion was hyper intense in T2 image and isointense in T1 image with no contrast enhancement in postgadolinium image.

**Surgery:** A C6-T1 laminoplasty was done under GA with prone position. Tumor was reached from Right side without significant cord retraction and removed with capsule. The cyst contains yellowish colored fluid. After removal of cyst the laminae was replaced with titanium mini screw and plate.

**Post-operative course:** Post operative recovery was uneventful. No new deficit was evident. From 1\textsuperscript{st} POD the symptoms was resolved dramatically. After 2 months a follow-up MRI was done which revealed no residual or recurrence.

**Histopathological examination:** revealed enterogenous cyst.

**Fig-1, 2, 3:** Pre-op MRI of cervical spine with contrast.

**Fig- 4:** Per-operative picture showing cyst removal.

**Fig- 5, 6, 7:** The boy at 2\textsuperscript{nd} POD. 3 months postoperative image showing no residual cysts.
Discussion:

Enterogenous cysts of the central nervous system, also called neuroenteric cysts or gastrocytomas, were first reported in 1934 by Pussep, who treated a case of intestinoma of the cervical spinal cord. These cysts within the spinal cord are not common. Using Table 1: Literature review

| Case | Author, year | Age(years) | Tumour location | Clinical | Surgery | Cyst content | Clinical outcome | Follow-up | Recurrence |
|------|--------------|------------|-----------------|----------|---------|--------------|-----------------|-----------|------------|
| 1    | Harriman DG, 1958 | 20/M       | T3, ID EM, D    | Chronic onset | LAM: T2-T4/PA | CSF-like fluid. | Died           | 1 year    | YES        |
| 2    | Pilz P et al, 1977 | 22/F       | C3-C4, ID EM, V | Acute onset | NO      | mucilage     | Died           | NR        | NO         |
| 3    | Mohanty S et al, 1979 | 23/F       | C5-C7, ID EM, V | Acute onset | LAM: C5-C7/PA | clear colourless fluid. | Improved     | 10 days   | NO         |
| 4    | Woe PY et al, 1982 | 1/M        | C2, ID EM, V    | Acute onset | LAM: C2 | clear colourless fluid | Improved     | NR        | NO         |
| 5    | Itohura T et al, 1986 | 4/F        | C1-C2, ID EM, D | Chronic onset | LAM: C1-C3/PA | CSF-like fluid | improved     | 9 months  | NO         |
| 6    | Aoki S et al, 1987 | 22/F       | C2-C3, ID EM, V | Chronic onset | LAM: C1-C4/PA | clear colourless fluid | Improved     | 4 weeks   | NO         |
| 7    | Lea ME et al, 1992 | 18/M       | C3-C7 ID EM, V  | Chronic onset | LAM: C3-C7/PA | NR           | Improved     | 7 days    | NO         |
| 8    | Chiang WH et al, 1992 | 5/M        | T5-T9, ID EM, V | Acute onset | PA      | milky white opalescent fluid | Improved     | NR        | NO         |
| 9    | Khandelwal N et al, 1993 | 25/M       | T9-T10, IM     | Chronic onset | LAM: T8-T11/PA | milky fluid | Improved     | 3 months  | NO         |
| 10   | Chen IH et al, 1995 | 30/M       | C7-T1, ID EM, V | Chronic onset | LAM: C6-T1/PA | NR           | improved     | 6 months  | NO         |
| 11   | Hamana-O T, 1997 | 7/M        | C4-C6, ID EM, D | Chronic onset | LAM: C5-C6/AA | NR           | Improved     | NR        | NO         |
| 12   | Lee SH et al, 1999 | 48/M       | T5-T6          | Chronic onset | COR: C5-C6/AA | NR           | Improved     | NR        | NO         |
| 13   | Shetty DS et al, 2000 | 3/M        | C7-T2, ID EM, V | Acute onset | LAM: C6-T3/PA | NR           | improved     | NR        | NR         |
| 14   | Reinders JW et al, 2001 | 35/F       | T8-T9, IM      | Chronic onset | LAM: T8-T9/PA | NR           | improved     | 2 months  | NO         |
| 15   | Martin AJ et al, 2001 | 35/F       | C7-T2, ID EM, V | Acute onset | LAM: T7-T8/PA | sterile, viscous, yellow fluid | Improved     | 7 months  | NO         |
| 16   | Chang IC, 2003 | 6/M        | C4-C6 ID EM, V | Chronic onset | LAM: C3-C7 | xanthochromic fluid | improved     | 3 years   | NO         |
| 17   | Hidennenzee T, 2003 | 3/F        | C2-C3, ID EM, V | Acute onset | LAM: C2-C4/PA | watery clear fluid | improved     | 5 years   | NO         |
| 18   | Shenoy SN, 2004 | 4/M        | C7-T1, ID EM, V | Acute onset | LAM: C2-C4/PA | watery clear fluid | poor         | 3 years   | NO         |
| 19   | 3/M        | C7-T1, ID EM, V | Acute onset | LAM: C2-C4/PA | watery clear fluid | improved     | 5 years   | NO         |
| 20   | 16/F       | C3-C4, ID EM, V | Chronic onset | LAM: C2-C4/PA | watery clear fluid | excellent | improved     | 3 years   | YES        |
| 21   | 5/F        | C6-T8, ID EM, D | Chronic onset | LAM: T6-T8/PA | milky, jelly-like fluid | improved     | 3 years   | YES        |
| 22   | Becker GW et al, 2004 | 59/F       | C3-C5, ID EM, V | Acute onset | LAM: C3-C5/AA | yellow keratinous material | maintain     | 6 months  | NO         |
| 23   | Arslan E et al, 2010 | 24/F       | L2, ID EM, D   | Chronic onset | LAM: T2-T4/PA | NR           | improved     | 9 months  | YES        |
| 24   | Zhu M et al, 2010 | 39/M       | T11-T12, IM    | Acute onset | LAM: T11-T12 | partially calcified | maintain     | NR        | NO         |
| 25   | Sadeghi-Hariri B et al, 2012 | 40/M | C1-L2, IM | Chronic onset | LAM: L1-L2/PA | creamy jelly-like contents | improved     | NR        | NO         |
| 26   | He ZG et al, 2015 | 8/M        | C7-T1, ID EM, V | Chronic onset | LAM: C6-T1/PA | NR           | Improved     | 4 months  | NO         |
| 27   | Can A et al, 2018 | 29/M       | C4-C7, ID EM, V | Chronic onset | LAM: C4-T1/PA | Mucinous transparent | Improved     | 7 months  | YES        |
| 28   | Yuce I et al, 2015 | 1/M        | T3-T4, ID EM, V | Acute onset | LAM: T3-T6/PA | NR           | Improved     | 3 months  | NO         |
| 29   | Jung HS, 2015 | 50/M       | T1, ID EM, V   | Acute onset | LAM: T1/PA | yellow-green mucinous fluid | improved     | 6 months  | NO         |
| 30   | Kejima S et al, 2016 | 2/M        | begin L1-L3, then L1-L2, ID EM, V | Acute onset | LAM: NR | watery clear fluid | improved     | 35 months | YES        |
| 31   | Joshi KC et al, 2017 | 8/M        | T3-T6, IM      | Chronic onset | LAM: T3-T6/PA | white pebble | Improved     | 3 months  | Ne         |

C, cervical; CSF, cerebrospinal fluid; COR, corpectomy; D, dorsal; ED, extradural; EM, extramedullary; F, female; ID, intradural; IM, intramedullary; L, lumbar; LAM, laminectomy; M, male; NR, not reported; PA, posterior approach; T, thoracic; V, ventral.
the keywords ‘neurenteric cyst’, ‘enterogenous cyst’, ‘spinal’ and ‘intraspinal’ on PubMed about 30 cases were reported, 6–34 including 10 women and 20 men, with a mean age of 22 years (range 1–59 years) with histological confirmation. They generally arise from the lower cervical to the upper thoracic region of the spinal cord, causing symptoms of spinal compression. Most of these cysts are found with intradural extramedullary location and intramedullary lesions are very rare. About half of these cases are associated with spinal deformities such as spina bifida, hemivertebra and vertebral fusion. Enterogenous cysts have been classified into three groups, according to histological features. Group A is the simplest type, lined by a single layer of cuboidal or columnar epithelial cells with or without cilia. Group B cysts include more complex elements of the gastrointestinal tract or tracheobronchial tree, including mucus glands and smooth muscle in their wall. Group C cysts have ependymal or glial tissue in addition to the elements seen in group B cysts. Most enterogenous cysts belong to group A. However, all our cases had features of group B.

A variety of hypotheses have been suggested regarding the pathogenesis of intraspinal enterogenous cysts, but none are firmly established. They are believed to originate from embryonal dysgenesis. During normal development, the neuroenteric canal closes and the notochord separates from the primitive gut in the third week of embryonic life. It is proposed that during the same period, a transient adhesion occurs between the neural ectoderm and endoderm, or a communication develops along the neuroenteric canal. When such a developmental abnormality persists because of the incomplete separation at this adherence or remnant canal, the cyst forms.

MacKenzie and Gilbert have demonstrated morphological and immunohistochemical similarities between colloid cysts of the third ventricle and spinal enterogenous cysts, suggesting that these lesions are all derived from primitive gut endoderm. Our case was a typical presentation with compressive cervical myelopathy with radiological and clinical presentation. MRI is the main tool of diagnosis. The lesion was extramedullary and intradural cystic lesion with elliptical shape located at the lower cervical and upper dorsal area which is its usual location. Other radiological presentation was typical.

Posterior approach was used for surgery rather than anterior. Laminoplasty of C6-T1 was done. Although some of the author has chosen anterior approach, we think posterior approach is enough for complete removal of the cyst.

Conclusion: Spinal enterogenous cysts are benign lesions with insidious progression, and their early preplanned surgical removal should be the goal of treatment, as the very advanced stage of manifestation can be critical for neural recovery and clinical outcome. Total resection is the first line treatment for patients with neurological impairment.

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