DOI: https://doi.org/10.3329/bjns.v11i1.57995

Case Report

Ventral Cervical Intradural Extramedullary Neuroenteric Cyst and its Operative Procedure; A Rare Case Report with Literature Review

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Received: 05 June, 2021
Accepted: 20 June, 2021

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 intraduralextramedullary 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 intraduralextramedullary 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.*

Conflict of Interest: *There is no conflict of interest relevant to this paper to disclose.*

Funding Agency: *Was not funded by any institute or any group.*

Bang. J Neurosurgery 2021; 11(1): 54-57

Introduction:

The intraspinalenterogenous cyst, also called aneurenteric 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[1]. These cysts result from inappropriate segmentation of the notochord during embryogenesis causing endodermal tissue to remain in the spinal canal[2]. It accounts for only 0.7-1.3% of spinal axis tumors[3].Only 12.2% of neuroenteric cysts aredocumented to be intramedullary[4]. They are benign epithelial lined cysts, with the lining resembling that of the alimentary canal[5]. They are often associated with developmental defects of the overlying skin and/or vertebral bodies[6]. Occasionally they can have fistulous connection with similar mediastinal, thoracic or abdominal cysts, thus supporting an endodermal origin of these cysts[7].

Such cysts were first reported by Kubie and Fulton in 1928 as teratomatous cysts,[8] and later described by Puusepp in 1934 as intestinomas[9]. Holcomb and

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

Case Report:

A 5 yrs boy presented with gradually progressive neck and upper back pain with quadriparesis 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 1st 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.

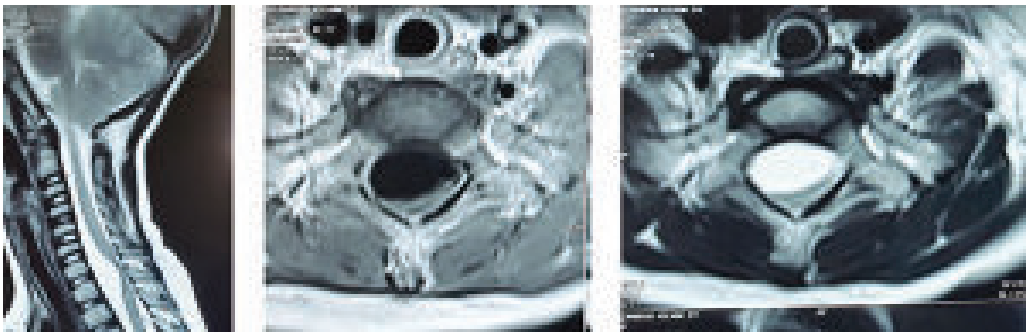

Fig- 1, 2, 3: *Pre-op MRI of cervical spine with contrast.*

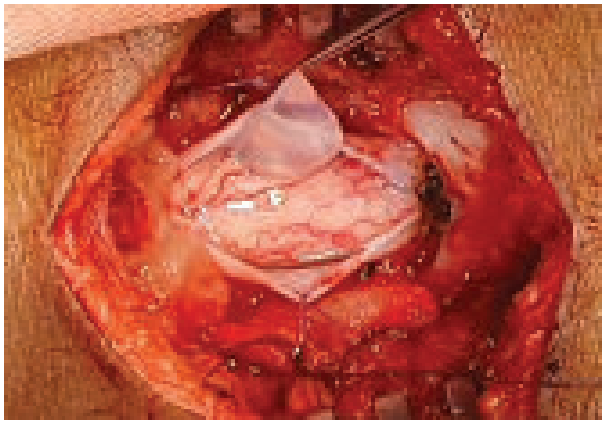

Fig.-4: *Per-operative picture showing cyst removal.*

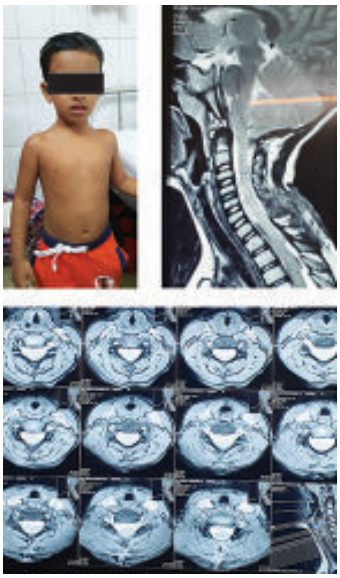

Fig- 5, 6, 7: *The boy at 2nd POD. 3 months postoperative image showing no residual cysts.*

Table-1: *Literature review*

Table 1 Cases of spinal enterogenous cysts.

Case	Author, year	Age(years) /sex	Tumour location	Clinical	Surgery	Cyst content	Clinical outcome	Follow-up	Recurrence
1	Harriman DG, 1958[6]	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[7]	22/F	C3–C4, ID EM, V	Acute onset	NO	mucilage	Died	NR	NO
3	Mohanty S *et al*, 1979[8]	23/F	C5–C7, ID EM, V	Acute onset	LAM: C5–C7/PA	clear colourless fluid.	Improved	10 days	NO
4	Woo PY *et al*, 1982[9]	1/M	C2, ID EM,V	Acute onset.	LAM: C2	clear colourless fluid	Improved	NR	NO
5	Itakura T *et al*, 1986[10]	4/F	C1–C2, ID EM, D	Chronic onset	LAM: C1–C3/PA	CSF-like fluid	excellent	9 months	NO
6	Aoki S *et al*, 1987[11]	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[12]	18/M	C3–C7 ID EM, V	Chronic onset	LAM: C3–C7/PA	NR	Improved	7 days	NO
8	Chhang WH *et al*, 1992[13]	5/M	T5–T9, ID EM, V	Acute onset	PA	milky white opalescent fluid	Improved	NR	NO
9	Khandelwal N *et al*, 1993[14]	25/M	T9–T10, IM,	Chronic onset	LAM: T8–T11/PA	milky fluid	Improved	3 months	NO
10	Chen IH *et al*, 1995[15]	30/M	C7–T1,ID EM, V	Chronic onset	LAM: C6–T1/PA	NR	Improved	6 months	NO
11	Hamamoto O, 1997[16]	7/M	C4–C6, D EM, D	Chronic onset	COR: C5–C6/AA	NR	Improved	NR	NO
12	Lee SH *et al*, 1999[17]	48/M	T5–T6	Chronic onset	NR	NR	Improved	NR	NO
13	Shetty DS *et al*, 2000[18]	3/M	C7–T2, ID EM, V	Acute onset	LAM: C6–T3/PA	NR	NR	NR	NR
14	Reinders JW *et al*, 2001[19]	35/F	T8–T9, IM	Chronic onset	LAM: T8–T9/PA	NR	Improved	2 months	NO
15	Martin AJ *et al*, 2001[20]	35/F	C7–T2, ID EM, V	Acute onset	COR: AA	sterile, viscous, yellow fluid	Improved	7 months	NO
16	Chang IC, 2003[21]	50/M	T7–T8, ID EM, V	Chronic onset	LAM: T7–T8/PA	viscous content	Improved	2 years	NO
17	Hicdonmez T, 2003[22]	6/M	C4–C6 ID EM, V	Chronic onset	LAM: C3–C7	xanthochromic fluid	Improved	3 years	NO
18	Shenoy SN, 2004[23]	4/M	C2–C3, ID EM, V	Acute onset	LAM: C2–C4/PA	watery clear fluid	Improved	5 years	NO
19		3/M	C7–T1, ID EM, V	Acute onset	LAM: C2–C4/PA	watery clear fluid	poor	3 years	NO
20		16/F	C3–C4, ID EM, V	Chronic onset	LAM: C2–C4/PA	watery clear fluid	excellent	3 years	NO
21		5/F	T6–T8, ID EM, D	Chronic onset	LAM: T6–T8/PA	milky, jelly-like fluid	Improved	3 years	YES
22	Becker GW *et al*, 2004[24]	59/F	C3–C5, ID EM, V	Acute onset	COR: C3–C5,AA	yellow keratinous material	maintain	6 month	NO
23	Arslan E *et al*, 2010[25]	24/F	L2, ID EM,D	Chronic onset	LAM: T2–T4/PA	NR	Improved	9 months	YES
24	Ziu M *et al*, 2010[26]	39/M	T11–T12, IM	Acute onset	LAM: T11–T12	partially calcified	maintain	NR	NO
25	Sadeghi-Hariri B *et al*, 2012[27]	40/M	L1–L2, IM	Chronic onset	LAM: L1–L2/PA	creamy jelly-like contents	Improved	NR	NO
26	He ZG *et al*, 2015[28]	8/M	C7–T1, ID EM, V	Chronic onset	LAM: C6–T1/PA	NR	Improved	4 months	NO
27	Can A *et al*, 2015[29]	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[30]	1/M	T3–T4, ID EM, V	Acute onset	LAM: NR	NR	Improved	NR	NO
29	Jung HS, 2015[31]	50/M	T1, ID EM, V	Acute onset	LAM: T1/PA	yellow-green mucinous fluid	improved	6 month	NO
30	Kojima S *et al*, 201632	2/M	begin L1–L3, then T12–L1, ID EM, V	Acute onset	LAM: NR	watery clear fluid	Improved	35 months	YES
31	Joshi KC *et al*, 2017[33]	8/M	T3–T6, IM	Chronic onset	LAM: T3–T6/PA	white pebble	Improved	3 months	No

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.

Discussion:

Enterogenous cysts of the central nervous system, also called neuroenteric cysts[10] or gastrocytomas[12], were first reported in 1934 by Pussep[9], who treated a case of intestinoma of the cervical spinal cord. These cysts within the spinal cord are not common, Using

the keywords 'neurenteric cyst', 'enterogenouscyst', 'spinal' and 'intraspinal' on PubMed about 30 caseswere reported,6–34 including 10 women and 20 men, with a mean age of 22 years (range 1–59 years) with histological confirmation[1].They generally arise from the lower cervical to the upper thoracic region of the spinal cord, causing symptoms of spinal compression[13]. Most of these cysts are found with intraduralextramedullary location and intramedullary lesions are very rare[14]. About half of these cases are associated with spinal deformities such as spina bifida, hemivertebra and vertebral fusion[13]. Enterogenous cysts have been classified into three groups, according to histological features[15]. 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 intraspinalenterogenous cysts, but none are firmly established[17]. They are believed to originate from embryonal dysgenesis[16]. 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[17].

MacKenzie and Gilbert[18] 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 it's usual location. Other radiological presentation was typical.

Posterior approach was used for surgery rather than anterior. LaminoplastryofC6-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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