A rare case of thoracic spinal intradural extramedullary enterogenous cyst with acute onset: case report and literature review

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ABSTRACT

The intraspinal enterogenous cyst, also called an neurenteric 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. Most patients present with symptoms of progressive focal pain, myelopathic signs or radicular symptoms. We report a rare case of thoracic spinal intradural extramedullary enterogenous cyst with rapidly progressive weakness of both lower extremities. Additionally, we analysed the literature concerning the clinical features, diagnosis and prognosis of this disease.

KEYWORDS

Enterogenous cyst – Intraspinal cyst – Neurenteric cyst – Spinal cord tumour

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Introduction

Intraspinal enterogenous cysts, also known as neurenteric cysts, are rare epithelium-lined structures of presumed endodermal origin.¹ Such cysts were first reported by Kubie and Fulton in 1928 as teratomatous cysts,² and later described by Puusepp in 1934 as intestinomas.³ Holcomb and Matson coined the term neurenteric cyst in 1954.⁴ The disease is officially named as enterogenous cyst in 1958 by Harriman.⁵ Previous studies of neurenteric cysts indicates that the disease is slowly progressive and rarely shows symptoms of acute onset. We report a case of a thoracic spinal intradural extramedullary enterogenous cyst with acute onset of bilateral lower extremity weakness. In addition, by searching the related literature using PubMed, we analysed the clinical features, diagnosis and prognosis of this disease.

Case history

Presentation

A 39-year-old woman presented with a 10-day history of back pain with zonaesthesia and progressive weakness of bilateral lower extremities, which had no specific cause. The pain was aggravated at night and did not alleviate after rest. Also, She had numbness and paraesthesia extending from her navel to her feet on both sides. Additionally, her medical history was notable for polycystic

ovary syndrome seven years ago, which was cured by surgery.

Examination

On physical examination, a bilateral lower extremity weakness was noted: power was grade manual muscle testing (MMT) 3/3 in both iliopsoas and 2/2 in the rest muscle groups in both legs. Her deep tendon reflexes were brisk in both lower limbs. The Babinski sign, patellar clonus and ankle clonus were present bilaterally. She had hyperreflexia of knees and ankles and patellar and ankle clonus, Babinski sign on both sides. Additionally, there was sensory loss below the belly button level but no sign of bladder and bowel involvement.

All laboratory tests were normal. Plain radiography of the thoracic spine indicated widening of the spinal canal at T9 and T10 and butterfly vertebra on the level of T4, T8 and L3. Chest x-ray showed no abnormality. Computed tomography (CT) without contrast material showed a hypodense mass at the level of T9 and T10 with a notably widening of the spinal canal. Moreover, an intraspinal extramedullary mass lesion which was compressing the cord at the level of thoracic T9–T10 was demonstrated on magnetic resonance imaging (MRI). It was isointense on T1-weighted images but hyperintense on T2-weighted imaging, and displayed no enhancement on contrast-enhanced MRI (fig 1A, B, C and D). Muscle strength of both lower limbs dropped to grade 0 at the third day of admission.

Figure 1 Intraoperative image showing cyst located in the ventral spinal cord, squeezing the spinal cord to the right, not adhering to the dura or the cord.

Figure 2 Histopathological examination revealing a cyst lined with pseudostratified ciliated columnar epithelium (haematoxylin and eosin staining $\times 200$).

Surgery

A T9–T10 laminotomy was performed in the prone position. With the help of a neurosurgical microscope, the spinal cord was visualised after durotomy and the mass located at the ventral side of spinal cord (fig 2). The mass lesion was cyst-like and contained jelly-like fluid. After opening the cyst, the fluid was removed by suction and the capsule wall was resected integrally after being carefully separated. The dura was continuously sutured and pedicle screw system was installed after thorough flushing. Intraoperative neurophysiological monitoring was stable during the operation.

Postoperative course

The patient showed remarkable improvement in the power of her lower limbs a few hours after the operation and the muscle strength returned to grade 4/4 in both legs on the fifth postoperative day. The patient could walk without assistance three months after surgery. No recurrence was found on postoperative MRI (fig 1 E and F).

Pathological examination

Histopathological examination revealed a cyst lined with pseudostratified ciliated columnar epithelium (fig 3) and immunohistochemistry showed CDX2 (–), CR (–), D2-40 (–), glial fibrillary acidic protein (–), napsin (–), S-100 (–), thyroid transcription factor-1 (–), villin (–). These results demonstrated that the mass lesion was an enterogenous cyst. She was eventually diagnosed with intraspinal enterogenous cysts.

Discussion

Intraspinal enterogenous cysts, also known as neurenteric cysts, account for 0.7–1.5% of all spinal cord tumours.⁶ The incidence of intraspinal enterogenous cysts is two to

one male to female, and it mostly occurs between the ages of 20 and 30 years. Enterogenous cysts only contain endodermal constituents, with epithelial tissues similar to those found in the digestive system. Cysts are generally considered to be related to residual or ectopic tissue embedded in the spinal cord and neuroectodermal during the third week of embryogenesis.⁷ Some patients also shows symptoms of congenital spinal deformity, such as vertebral body fusion, butterfly vertebra, spinal bifida, skin fistula and scoliosis.

Using 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) (Table 1). The lesions were localised to C1 to L2, with the majority located in the cervicothoracic region, and rarely seen in the lumbar spine and cauda equine.^{27,32} Among these cases, there were 27 intradural extramedullary enterogenous cysts; 22 lesions were located at the ventral side of the cord; only 3 were located in the cord. In these cases, most patients presented progressive focal pain at the level of the spine, various myelopathic signs or radicular symptoms, such as quadriplegia, chronic pyrexia, neck pain, sphincter incontinence and paraplegia. In addition, other symptoms including loose spontaneous respiration, urinary incontinence and diminished temperature sensation.

MRI is the main tool for diagnosis in the patient with signs or symptoms associated with intraspinal enterogenous cysts. The most common findings are isointense on T1-weighted images and hyperintense on T2-weighted imaging and display no enhancement on contrast-enhanced MRI. The cord develops faster than the lesion and gradually coats the cyst during the process of growth. This leads to the typical ‘spinal-cord-insertion’ sign on

Figure 3 T1-weighted image showing isointense lesion (A); T2-weighted image showing hyperintense lesion (B); T4, T8 and L3 level are butterfly vertebra (C and red arrow); no enhancement on contrast-enhanced magnetic resonance imaging (D); no recurrence was found on postoperative magnetic resonance imaging three months later (E, F).

MRI, which suggests that it may be a congenital disease lesion. In some intramedullary cases, the mass could be partially calcified. CT plays an important role in the evaluation of the calcification and spinal deformities associated with enterogenous cysts.

The epithelial tissues of cyst are similar to the digestive system histopathologically. The cyst wall linings range from a simple cuboidal epithelial layer to columnar epithelial layer and some reports show that a pseudostratified epithelial layer resting on a thin layer of fibrous connective tissue may also present in the cyst. In addition, Goblet cells and ciliated cells are commonly observed in varying proportions of the cysts.

Enterogenous cysts were classified into three types by Wilkins and Odom.³⁴ Type A includes cysts containing either columnar or cuboidal cells, with ciliated and non-ciliated components atop a basal membrane composed of type IV collagen. Type B includes cysts with all of the features of type A as well as additional tissue that may include bone, cartilage, lymphatic tissue, fat or glandular components. Type C cysts are identified by type A features in

association with ependymal original tissue. Some 90% of enterogenous cysts are type A.

Surgical treatment is considered the main therapy for patients with sensory and motor deficits associated with enterogenous cysts. Most symptoms can be significantly alleviated, except in a few cases where the cyst recurred or was exacerbated. Laminotomy is the most frequently and safely used approach for cases with lesion located to the dorsal and ventral of the cord. Corpectomy though anterior approach can be used in cervical lesions.^{55,57} Reports of postsurgical recurrence have ranged between 0% and 37%.⁵⁶

Conclusions

The enterogenous cyst is a rare congenital spinal tumour. Most patients present with symptoms of progressive focal pain, myelopathic signs or radicular symptoms acutely or chronically. MRI is the first choice for diagnosis. Total resection is the first-line treatment for patients with neurological impairment.

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 <i>et al</i> , 1977 ⁷	22/F	C3–C4, ID EM, V	Acute onset	NO	mucilage	Died	NR	NO
3	Mohanty S <i>et al</i> , 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 <i>et al</i> , 1982 ⁹	1/M	C2, ID EM, V	Acute onset.	LAM: C2	clear colourless fluid	Improved	NR	NO
5	Itakura T <i>et al</i> , 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 <i>et al</i> , 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 <i>et al</i> , 1992 ¹²	18/M	C3–C7 ID EM, V	Chronic onset	LAM: C3–C7/PA	NR	Improved	7 days	NO
8	Chhang WH <i>et al</i> , 1992 ¹³	5/M	T5–T9, ID EM, V	Acute onset	PA	milky white opalescent fluid	Improved	NR	NO
9	Khandelwal N <i>et al</i> , 1993 ¹⁴	25/M	T9–T10, IM,	Chronic onset	LAM: T8–T11/PA	milky fluid	Improved	3 months	NO
10	Chen IH <i>et al</i> , 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 <i>et al</i> , 1999 ¹⁷	48/M	T5–T6	Chronic onset	NR	NR	Improved	NR	NO
13	Shetty DS <i>et al</i> , 2000 ¹⁸	3/M	C7–T2, ID EM, V	Acute onset	LAM: C6–T3/PA	NR	NR	NR	NR
14	Reinders JW <i>et al</i> , 2001 ¹⁹	35/F	T8–T9, IM	Chronic onset	LAM: T8–T9/PA	NR	Improved	2 months	NO
15	Martin AJ <i>et al</i> , 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 <i>et al</i> , 2004 ²⁴	59/F	C3–C5, ID EM, V	Acute onset	COR: C3–C5,AA	yellow keratinous material	maintain	6 month	NO
23	Arsian E <i>et al</i> , 2010 ²⁵	24/F	L2, ID EM,D	Chronic onset	LAM: T2–T4/PA	NR	Improved	9 months	YES
24	Ziu M <i>et al</i> , 2010 ²⁶	39/M	T11–T12, IM	Acute onset	LAM: T11–T12	partially calcified	maintain	NR	NO
25	Sadeghi-Hariri B <i>et al</i> , 2012 ²⁷	40/M	L1–L2, IM	Chronic onset	LAM: L1–L2/PA	creamy jelly-like contents	Improved	NR	NO
26	He ZG <i>et al</i> , 2015 ²⁸	8/M	C7–T1, ID EM, V	Chronic onset	LAM: C6–T1/PA	NR	Improved	4 months	NO
27	Can A <i>et al</i> , 2015 ²⁹	29/M	C4–C7, ID EM, V	Chronic onset	LAM: C4–T1/PA	Mucinous transparent	Improved	7 months	YES
28	Yuce I <i>et al</i> , 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 <i>et al</i> , 2016 ³²	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 <i>et al</i> , 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.

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