



CASE REPORT

Intramedullary endodermal cyst including glial tissues in the spinal cord

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Received: 26 February 2020 / Revised: 19 April 2020 / Accepted: 21 April 2020
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Abstract

Introduction Spinal intramedullary endodermal cyst is a rare spinal cord tumour. In particular, an endodermal cyst that includes glial tissues is extremely rare. Herein, we present the case of an individual with a thoracic spinal cord intramedullary endodermal cyst, which includes glial tissues that achieved gross total resection by surgery.

Case presentation A 59-year-old man presented with a 10-month history of right thigh pain and numbness. Magnetic resonance imaging (MRI) revealed a well-margined 15-mm cystic lesion at the T7–T8 level. We performed cystectomy and achieved gross total resection. Pathological findings revealed an endodermal cyst, with the presence of glial tissues. No recurrence of cysts was observed upon MRI 2 years after the surgery.

Discussion Endodermal cyst is defined by pathological findings of a cyst lined by columnar epithelium of presumed endodermal derivation. To date, only 104 reported cases of intramedullary endodermal cysts have been reported; our report was the third case that showed the presence of glial cells in the cyst during pathological examination. Intramedullary cysts are generally difficult to completely resect, with many recurrences. Although we achieved gross total resection, careful follow-up is necessary in the future.

Introduction

Endodermal cysts are rare lesions accounting for 0.7–1.3% of all spinal cord tumours. Approximately 90% of these tumours are located in the intradural extramedullary compartment, whereas the remaining 10% are in the intramedullary compartment [1]. In particular, an endodermal cyst including glial tissues is extremely rare. Herein, we present the case of an individual with a thoracic spinal cord intramedullary endodermal cyst with the presence of glial tissues that achieved gross total resection.

Case presentation

A 59-year-old man presented with a 10-month history of right thigh pain and numbness. However, his medical history was unremarkable.

Physical examination revealed right thigh pain and numbness, with sensory disorder of the lower half of the body. He had exaggerated bilateral patellar tendon and Achilles tendon reflexes and positive bilateral Babinski reflexes. However, his lower limb strength and bowel and bladder functions were normal. The Japan Orthopaedic Association (JOA) score is a scoring system for thoracic myelopathy that evaluates the motor and sensory functions of the lower extremity and bladder function, and has a total of 11 points. In this case, the JOA was 7.5/11.

X-ray and computed tomography revealed no structural anomalies. MRI revealed the presence of a well-margined 15-mm cystic lesion with hypointensity on T1-weighted image, hyperintensity on T2-weighted image and a solid component at the T7–T8 level. The wall of the cyst was slightly enhanced on contrast T1-weighted image (Fig. 1). Ependymal cysts, dermoid cysts, epidermoid cysts and endodermal cysts were suspected as intramedullary cystic

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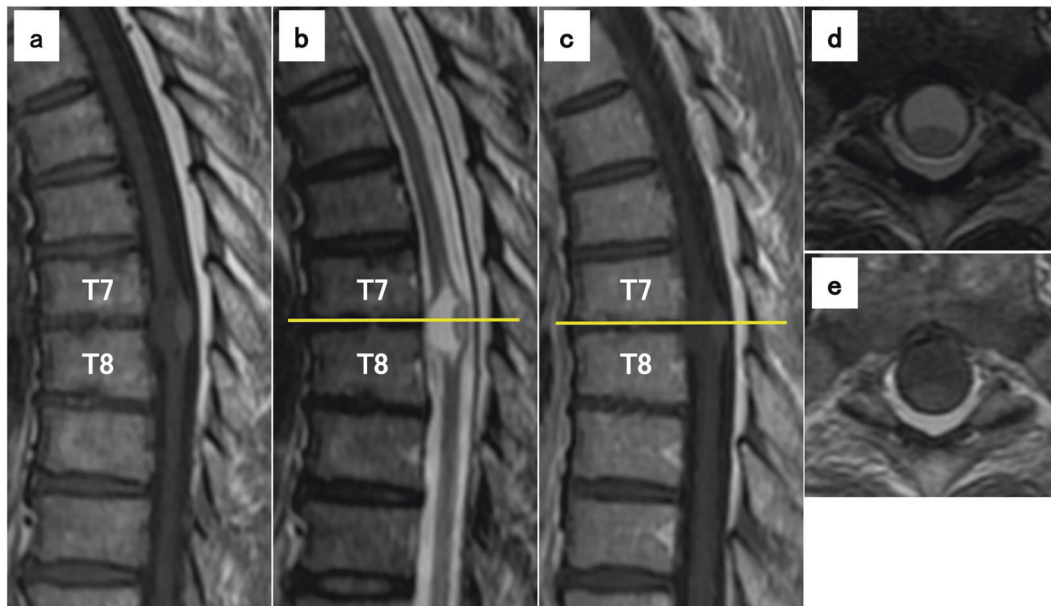
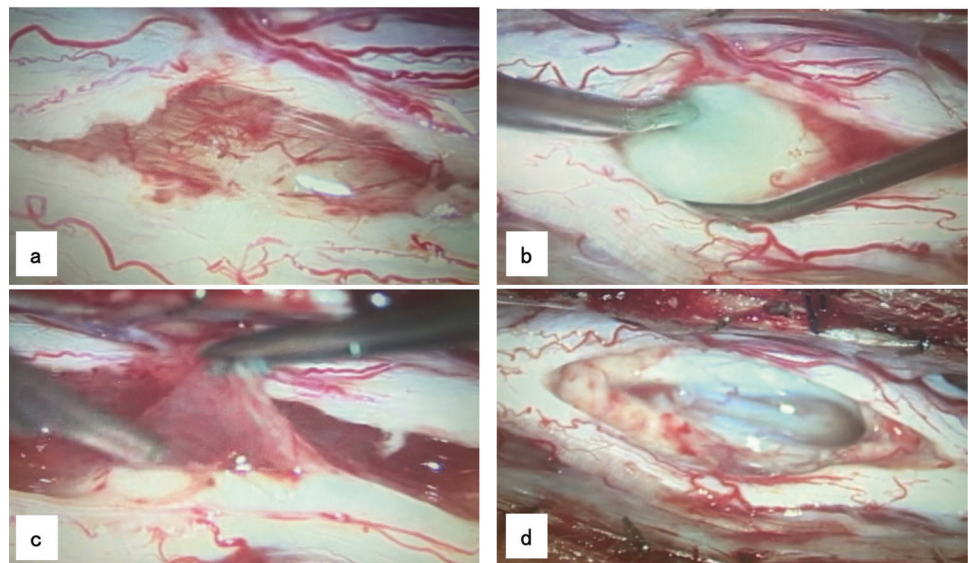


Fig. 1 Preoperative magnetic resonance imaging. **a** Sagittal T1; **b** sagittal T2; **c** sagittal Gd; **d** axial T2; **e** axial Gd.

Fig. 2 Intraoperative findings.

a Breaking of the posterior median sulcus; **b** incision of the capsule of the cyst; **c** removal of the capsule; **d** achievement of gross total resection.



lesions; however, the image could not provide a clear distinction. Considering the diagnostic findings, we performed cystectomy.

First, we performed laminectomy from T7 to T9 and observed the interior part of the dura using ultrasonography to confirm the localisation of the cyst. Subsequently, we incised the dura and approached the cyst by breaking the posterior median sulcus and exposed the cyst capsule using an intraoperative microscope. On incising the capsule, we observed the accumulation of a white turbid liquid. We removed this liquid and excised the capsule from the normal spinal cord whilst closely monitoring the motor evoked potentials. We performed this procedure with utmost care

because the capsule adhered to the normal spinal cord, achieving gross total resection of the cyst (Fig. 2).

In the cyst, we observed lining rows of ciliated columnar epithelial cells that were positive for pan-cytokeratin staining, an epithelial marker. The individual was then diagnosed with an endodermal cyst. Glial fibrillary acidic protein (GFAP)- and S100-positive glial cells were also observed in the subepithelial connective tissue (Fig. 3). According to Wilkins and Odom classification, the cyst was a type C cyst, which includes glial tissues [2].

His right thigh pain has improved since 2 years, with a JOA score of 7.5/11. Furthermore, MRI revealed no recurrence of the cysts (Fig. 4).

Fig. 3 Pathological findings. **a** The cyst is lined with ciliated columnar epithelial cells (arrows), H&E; **b** positive for pan-cytokeratin staining; **c** positive for GFAP; **d** positive for S100.

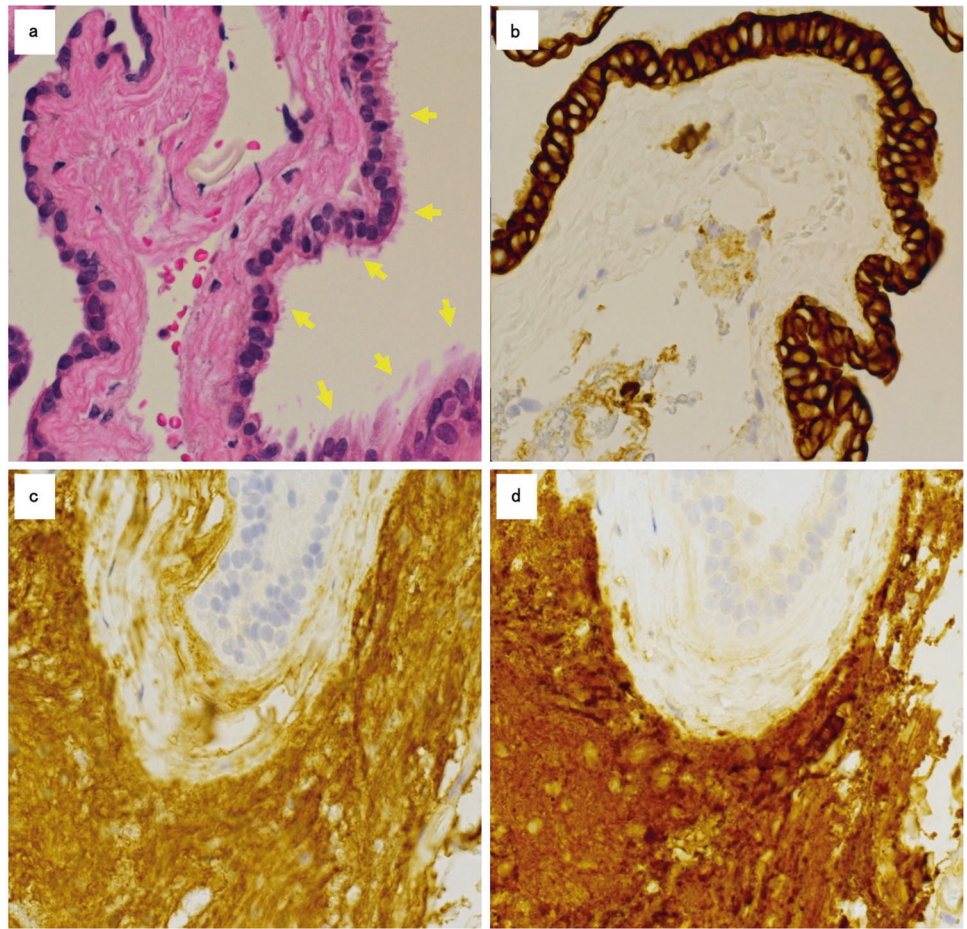
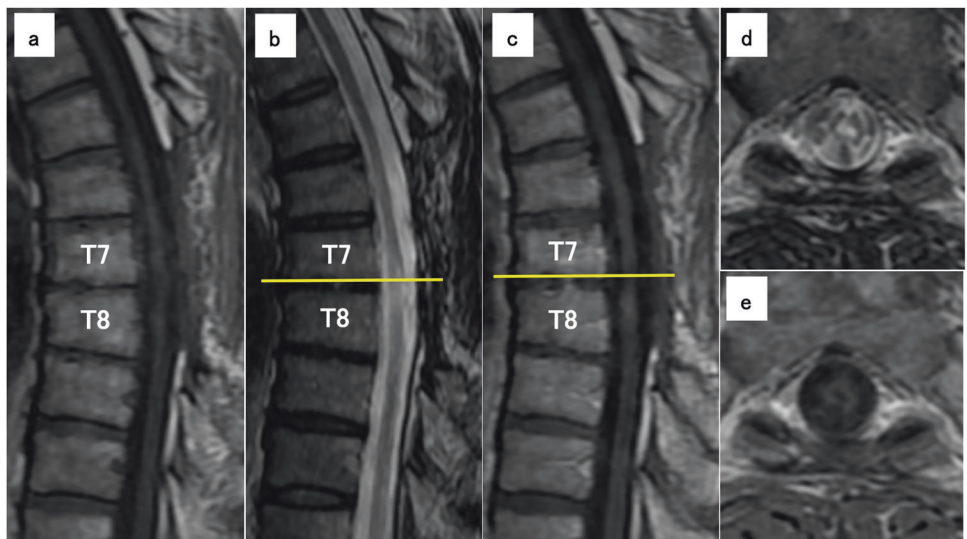


Fig. 4 Magnetic resonance imaging 2 years after the surgery. **a** sagittal T1; **b** sagittal T2; **c** sagittal Gd; **d** axial T2; **e** axial Gd.



Discussion

The precise pathogenesis of endodermal cysts still remains unclear. One of the most widely accepted aetiologies of cysts is the failure of separation of the ectoderm (notochord)

and the endoderm (foregut) during the third week of human embryogenesis [3]. The frequency with which it occurs in males and females is ~2:1 [4]. Endodermal cyst diagnosis is based on pathological findings. Endodermal cysts are defined as cysts lined by columnar epithelium of presumed

Table 1 Summary of intramedullary endodermal cysts described in the literature.

Year	Author	Age	Sex	Cyst location	Treatment	Recurrence, re-growth or refilling	FU (month)
1972	Silvernail and Brown	12 years	M	C2-T2	STR and Shunting	No	16
1988	Mizuno et al.	6 years	M	C2-5	Aspiration	Yes	96
1993	Khandelwal et al.	25 years	M	T9-10	GTR	No	3
1996	Rao et al.	27 years	M	C7	STR	No	30
1997	Rivierez et al.	46 years	F	L1-2	Shunting	No	6
2001	Lippman et al.	68 years	F	T10-11	STR	No	1.5
2001	Reinders et al.	3 years	F	T8-9	STR	No	2
2001	Sharma et al.	24 years	F	L1-2	STR	Yes	60
2002	Kumar and Nayak	9 years	F	C5-T1	STR	No	6
		15 months	M	L2-4	STR	No	3
2003	Paolini et al.	28 years	F	T8-9	STR	No	12
2005	de Oliveira et al.	6 years	M	C4-7	GTR	Yes	96
		6 years	F	C7-T1	GTR	No	168
		21 days	M	T10	GTR	No	0.75
2005	Rotondo et al.	67 years	F	T10-11	GTR	No	3
		53 years	F	T12-L1	GTR	No	12
		61 years	F	T12-L1	GTR	No	6
2006	Menezes and Traynelis	6 years	M	C2-3	STR	Yes	216
		40 years	M	C6	STR	Yes	36
2007	Nagi et al.	40 years	F	C3-4	GTR	No	3
2008	Cai et al.	3 years	F	C7-T1	GTR	No	60
2008	Muzumdar et al.	12 years	M	C2-3	GTR	No	36
2008	Takahashi et al.	8 years	M	T5	Aspiration	Yes	36
2008	Garg et al.	9 years	F	T1	STR	Yes	36
		10 years	M	T3-5	STR	No	12
		14 years	M	C3	GTR	No	84
		22 years	F	C7-T2	STR	No	36
		25 years	M	C5-7	GTR	No	2
2009	Yilmaz et al.	17 years	M	T12-L1	STR	No	6
2010	Theret et al.	30 days	M	C6-7	STR	No	24
2011	Balasubramaniam et al.	2 years	M	T12-L1	STR	No	12
2011	Wakao et al.	57 years	M	T1-2	STR and Shunting	No	12
2015	Tao et al.	10 years	M	C3-5	GTR	No	122
		25 years	M	L3	GTR	No	114
		53 years	F	T11-12	GTR	No	102
		43 years	M	C3-6	STR	Yes	93
		39 years	M	T5	GTR	No	84
		41 years	F	C3-4	STR	Yes	70
		47 years	M	L1	STR	Yes	65
		64 years	M	T7	GTR	No	54
		23 years	M	C3-4	GTR	No	50
		13 years	F	T1-2	STR	Yes	42
		15 years	F	C6-T1	GTR	No	34
		53 years	F	T10-12	GTR	No	23
		51 years	F	C5	STR	No	11
2016	Bruzek et al.	47 years	M	C2-C7	GTR	No	3
2017	Aiyappan et al.	34 years	M	T12-L1	STR	No	24
2017	Joshi et al.	8 years	M	T3-6	GTR	No	3
2019	Chen et al.	46 years	F	C0-1	STR	No	3

endodermal derivation [5]. Cysts with these features have been variably designated as neuroenteric, enterogenous or bronchogenic. The term ‘endodermal’ encompasses the

lesions occurring intracranially and throughout the spinal axis, as defined by the Armed Forces Institute of Pathology (AFIP) in 1993 [6]. Wilkins and Odom have pathologically

classified endodermal cysts into three types. Type A cysts compose of columnar or cuboidal cells atop a basement membrane, without other elements. Type B cysts include all of the features of type A and additional tissue that may include bone, cartilage, lymphatic tissue, fat, or glandular components, such as respiratory or intestinal organs. Type C cysts have type A features associated with ependymal or glial tissues [2, 4]. In the present case, the cyst was lined with ciliated columnar epithelial cells and was positive for pan-cytokeratin staining, indicating the typical diagnosis of an endodermal cyst. Furthermore, this case showed the presence of glial cells that were positive for GFAP and S100 in the subepithelial connective tissue, suggesting the cyst to be a type C cyst. Endodermal cysts are typically negative for GFAP; however, some studies have reported that cysts may be positive for GFAP in some intramedullary cases [2, 4, 7]; our case is consistent with these reports. To the best of our knowledge, our case is the third case of type C cyst of intramedullary endodermal cyst [2, 7].

In the present case, MRI revealed the presence of a well-marginated cystic lesion with hypointensity on T1-weighted image, hyperintensity on T2-weighted image and slightly enhanced wall on contrast T1-weighted image. These findings are typical of various cystic lesions, including endodermal cysts. Therefore, differentiating our individual's cyst from other cystic lesions based on MRI findings alone was difficult. Previous MRI findings of endodermal cysts, such as being well-marginated, hypointense on T1-weighted image, hyperintense on T2-weighted image and slightly enhanced wall on contrast T1-weighted image, are similar to those of our case [8, 9]. However, a characteristic finding of endodermal cysts is that the content fluid comprises mucin and protein components; therefore, various signal intensities can be exhibited depending on their properties, viscosity, and concentration [8, 9].

Ependymal cysts can be differentially diagnosed as spinal intramedullary cystic lesions; however, as described above, it is difficult to diagnose based on the image findings alone. Ependymal cysts are defined as appropriate for benign, intraparenchymal and often paraventricular cysts in which the lining lacks cilia and shows the presence of S100 protein and occasionally GFAP-reactive cells [5], which distinguishes it from endodermal cysts.

Endodermal cysts are benign lesions; however, the primary pathogenic mechanism of intramedullary lesions is chronic spinal cord compression. Therefore, surgery is the first treatment choice for neural decompression and prevention of cystic refilling [10]. In addition to resection, cyst aspiration and cyst-subarachnoid shunting have been performed. However, aspiration cannot yield a pathological diagnosis and results in a high refilling rate [1, 10–15]. The spillage of cyst contents into the subarachnoid space caused

Table 2 The surgical method, postoperative recurrence, and observation period of 49 previous cases.

Method	Cases	Recurrence, re-growth or refilling	Observation period
GTR	22	1 (4.5%)	48.3 months
STR	22	8 (36.4%)	36.4 months
Aspiration	2	2 (100%)	66.0 months
Shunting	3	0 (0.0%)	11.3 months

GTR gross total resection, STR subtotal resection.

during shunting may cause meningitis [1, 15]. Therefore, we performed resection to avoid these risks.

Our extensive research revealed that only 104 cases of intramedullary endodermal cysts have been reported. Of these, 49 confirmed the surgical method, postoperative recurrence, re-growth or refilling and observation period (Table 1). In addition, 22 cases achieved gross total resection, whereas another 22 cases achieved subtotal resection because the cyst was adhered to the normal spinal cord. Two cases underwent aspiration and three cases underwent shunting. We found one case of recurrence (4.5%) after gross total resection and eight cases of re-growths (36.4%) after subtotal resection (Table 2). Therefore, achieving gross total resection reduces the risk of recurrence, re-growth or refilling and is an effective treatment for cysts.

In the present case, we carefully removed the capsule of the cyst from the normal spinal cord. No recurrence has been observed 2 years after the achievement of gross total resection. Previous studies have reported that the possibility of recurrence in this case may not be high; however, reports on the long-term outcomes of intramedullary endodermal cysts achieving gross total resection are still few; thus, careful follow-up is required.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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