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Case Report

Intramedullary neurenteric cyst associated with a tethered spinal cord: Case report and literature review

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Abstract

Background: Neurenteric cysts are benign tumors of the central nervous system (CNS) that represent 0.3% to 0.5% of all spinal cord tumors. They are usually extramedullary and found in the lower cervical and thoracic spine. Only 12.2% of neurenteric cysts are documented to be intramedullary.

Case Description: The authors report a case of a 35-year-old female that presented with progressive weakness and loss of coordination in her legs. Magnetic resonance imaging (MRI) showed an intramedullary cystic lesion in the thoracolumbar region and a low-lying conus medullaris suggesting tethered cord. The patient was taken to the operating room for detethering of her spinal cord and resection of the lesion. Pathologic examination of the tissue confirmed the diagnosis of a neurenteric cyst.

Conclusion: A search of the literature since the advent of MRI showed 29 published cases of intramedullary neurentic cysts. Of the 24 published cases with a follow-up MRI, the average recurrence rate was 25% with a mean follow up of 51 months.

Key Words: Intramedullary, neurenteric cyst, tethered spinal cord

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INTRODUCTION

Neurenteric cysts, also known as enterogenous cysts or endodermal cysts, are benign tumors of the central nervous system (CNS) which are thought to represent 0.3% to 1.3% of all spinal cord tumors.[15,23] They are the result of inappropriate partitioning of the embryonic notocordal plate and presumptive endoderm during the third week of human development.[23] The cysts are lined by nonciliated epithelium that is simple or pseudostratified and cuboidal or columnar.[5] They are found to have a male predominance (60.4%) and present at a mean age of 6.4 years in the pediatric population[6] and in the second or third decades of life in the adult population.[23] The embryological origin is thought to be from an abnormal communication between the primitive neurenteric canal, notocord, and neural tube to the adjacent endoderm and mesenchyme during the third week of embryogenesis.[15]

Since the advent of magnetic resonance imaging (MRI), it has become possible to diagnose these lesions preoperatively. On T2-weighted imaging, the cysts are hyperintense and display minimal or no enhancement on T1 postcontrast imaging. Typically, the cysts are intradural/extramedullary (78.6%) and usually arise from the cervical, cervicothoraric, and thoracic spine (73.6%).[6]

This case is unique because the patient presented at the age of 35 with an intramedullary neurenteric cyst in the thoracolumbar spine and a tethered spinal cord. In

this paper, we will discuss the treatment of this patient as well as a review of all published literature on patients with intramedullary neurenteric cysts since the advent of MRI.

CASE REPORT

Presentation

A 35-year-old female presented with weakness in both legs and loss of coordination over the last year. She had some numbness and burning from her feet to her knees bilaterally and intermittent episodes of bladder incontinence.

Examination

On examination, she had no cranial nerve deficits and strength and sensation were normal in both upper extremities. She had bilateral lower extremity weakness, grade 3/5 in both dorsiflexors, and 4/5 in all other muscle groups. Her reflexes were 1+ at the knees and ankles with a very wide-based antalgic gait.

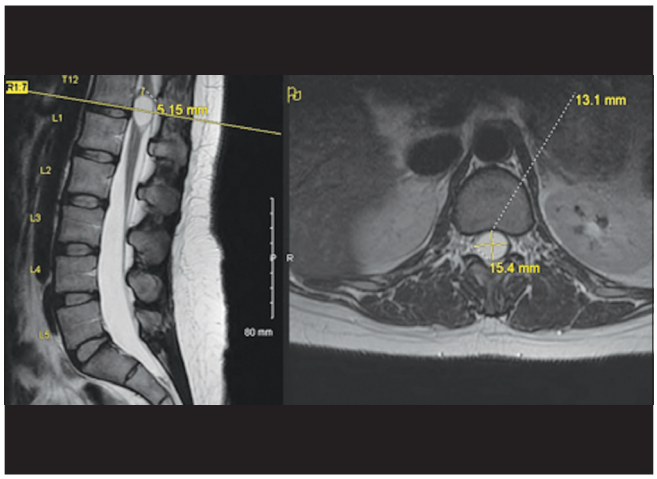

Figure 1:T2 sagittal and axial

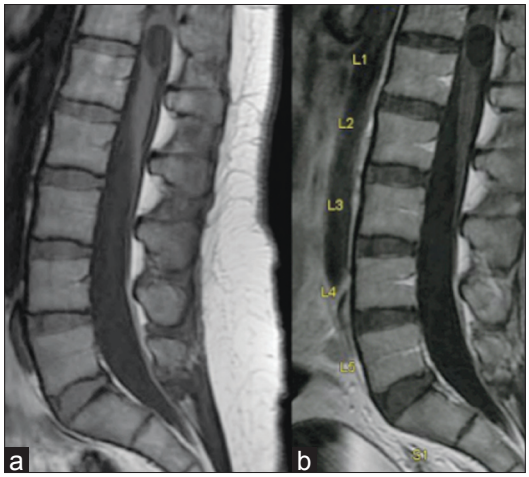

Figure 2:T1 sagittal precontrast (a) and postcontrast (b)

Imaging characteristics

MRI of the spine was performed which showed a cystic dilation of her distal cord near T12-L1 that had a similar intensity to cerebrospinal fluid (CSF) and no contrast enhancement. The cord also appeared to be low lying and terminated at the level of L3 [Figures 1 and 2].

Operation

A laminectomy was performed from T11 to L1 and L5 to S1. Initial attention was focused at the L5-S1 level where an intradural dissection was performed and the filum terminale was indentified, cauterized, and sectioned. After release of the tethered cord, the dura was opened from T11 to L1 and the spinal cord was visualized with an intraoperative microscope. A midline myelotomy was performed and a firm capsule was encountered within the cord. The cyst capsule was incised to internally decompress the cyst and allow for resection while minimizing trauma to the normal surrounding spinal cord. A milky white fluid was expressed upon opening the cyst. Microsurgical resection of the cyst capsule was performed while closely monitoring motor evoked potentials (MEP) and somatosenory evoked potentials (SSEP). A large portion of the cyst wall was resected, but a complete resection was aborted after an 80% decrease in MEP.

Postoperative course

The patient awoke with increased weakness, in comparison to her preoperative exam, with 2/5 hip flexors, 3/5 knee extensors, 1/5 knee flexors, and absent dorsiflexors, and plantar flexors. Her neurological function gradually improved over the next four weeks. With rehabilitation, her strength and coordination improved with the exception of her left dorsiflexors, which remained 3/5 strength.

Pathological examination

Hematoxylin and eosin staining revealed multiple irregular cystic spaces lined by epithelium [Figure 3]. Most areas showed simple or stratified cuboidal cells, but some areas had a squamous appearance. Immunohistochemical stains were reactive for pankeratin,

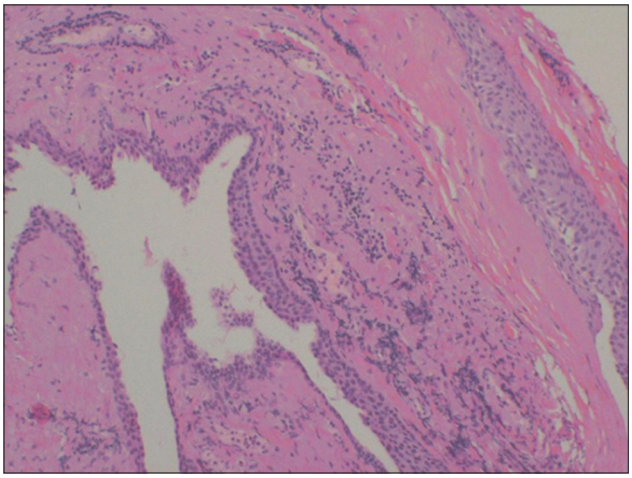

Figure 3: Hematoxylin and eosin stain

cytokeratin-7, epithelial membrane antigen, and carcinoembryonic antigen [Figure 4]. The epithelium was nonreactive for cytokeratin-20, thyroid transcription factor-1, and glial fibrillary acidic protein [Figure 5]. The surrounding neuroglial tissue shows strong reactivity for glial fibrillary acidic protein.

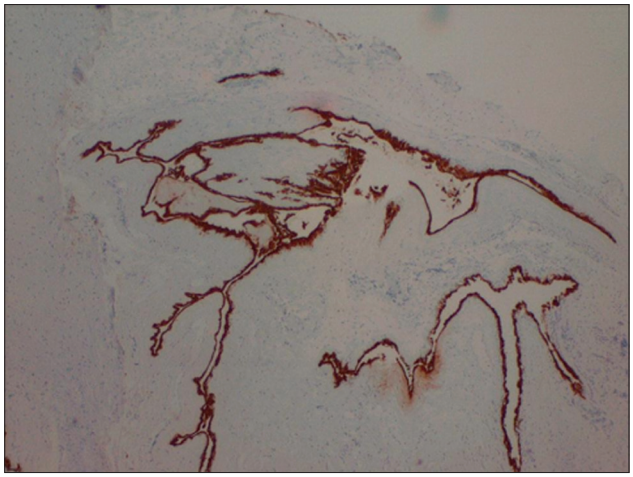

Figure 4: Cytokeratin 7 stain

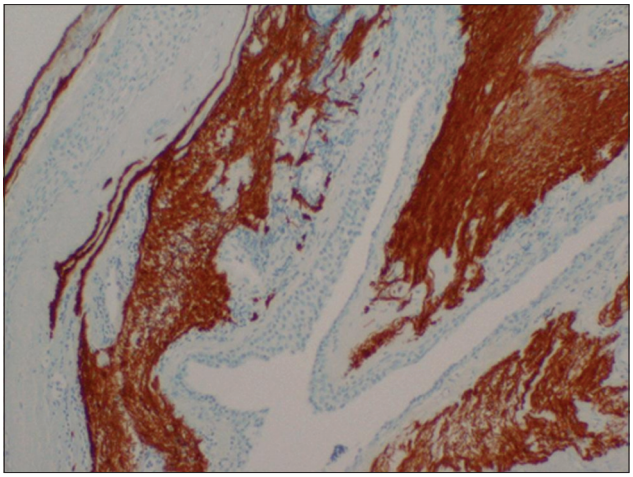

Figure 5: Glial fibrillary acidic protein stain

Table 1: Published cases of intradural neurenteric cysts[1,2,10,11,14-22,24,25,27,28,30,31]

Source	Age	Sex	Location	Treatment	Recurrence on MRI	Follow-up (months)	New deficits on last follow-up
Rivierez (1997)	46	F	L1-L2	Biopsy and cysto-arachnoid shunt	No	6	Yes
Reinders (2001)	31	F	T8-T9	Subtotal resection	No	2	Yes
Rauzzino (2001)	19	F	T12-L2	Subtotal resection	Yes	72	No
Rauzzino (2001)	6 wks	F	T12-L2	Subtotal resection	No	84	No
Sharma (2001)	24	F	L1-L2	Subtotal resection	Yes	60	No
Singal (2001)	67	M	T7	Total resection	Unknown	Unknown	Unknown
Lippman (2001)	68	F	T10-T11	Subtotal resection	No	1.5	No
Agrawal (2002)	3 mon	M	T1-T7	Total resection	Unknown	Unknown	No
Paolini (2002)	28	F	T8-T9	Subtotal resection	No	12	No
De Oliveira (2005)	6	M	C4-C7	Total resection	Yes	96	No
De Oliveira (2005)	6	F	C7-T1	Total resection	No	168	No
De Oliveira (2005)	3 wks	M	T10	Total resection	No	0.75	Deceased
Rotondo (2005)	67	F	T10-T11	Total resection	No	3	No
Rotondo (2005)	52	F	T12-L1	Total resection	No	12	No
Rotondo (2005)	61	F	T12-L1	Total resection	No	6	No
Menezes (2006)	6	M	C2-C3	Subtotal resection	Yes	216	No
Menezes (2006)	4	F	C4-C6	Total resection	No	156	No
Menezes (2006)	40	M	C6	Subtotal resection	Yes	36	No
Nagi (2007)	40	F	C3-C4	Subtotal resection	No	3	No
Muzumdar (2008)	12	M	C2-C3	Total resection	No	36	No
Cai (2008)	3	F	C7-T1	Subtotal resection	No	60	No
Takahashi (2008)	8	M	T5	Biopsy and aspiration	Yes	36	No
Yilmaz (2009)	17	M	T12-L1	Subtotal resection	No	6	No
Aydin (2009)	14 wks	F	C4-T1	Total resection	No	120	No
Theret (2010)	4 wks	M	C6-7	Subtotal resection	No	24	No
Ziu (2010)	39	M	T11-T12	Total resection	Unknown	Unknown	Unknown
Kleklamp (2011)	50	M	C6-T1	Total resection	Unknown	Unknown	No
Jhawar (2012)	3	M	C6-T3	Subtotal resection	Unknown	12	No
Present case	35	F	T12-L1	Subtotal resection	No	5	No

DISCUSSION

Intramedullary neurenteric cysts are thought to represent 12.2% of all neurenteric cysts. In the published pediatric literature, the cervical spine was the most common presenting location (36%).[6] In our review, we found the lower thoracic and thoracolumbar spine to be the most common location for intramedullary neurenteric cysts.

The treatment options for intramedullary neurenteric cysts remain controversial. Most authors advocate total resection when possible.[1,5,15] Menezes and Traynelis recommend an anterior approach because the lesions are typically ventral to the spinal cord.[15] De Oliveira, on the other hand, recommends a posterior approach because it is technically easier and causes fewer complications for the patient.[5] Takahashi has published his experience with MR imaging-guided, percutaneous aspiration and believes it should be the treatment of choice, with the caveat that conventional surgery may be necessary in the future.[27]

In the current published literature, the overall recurrence rate for neurenteric cysts after surgical resection varies from 11.6% to 37%.[6,23] To our knowledge, there is no published data on the recurrence rates of intramedullary neurenteric cysts. Of the 24 published cases with a follow-up MRI [Table 1], six (25%) showed radiographic signs of recurrence at a mean follow up of 51 months. Four underwent a subtotal resection, one a total resection, and one a MRI-guided percutaneous aspiration. None of the patients that had a gross total resection of their cyst had a recurrence. There was one death due the respiratory complications in a 3-week-old and two patients who had new deficits on their last follow-up visit.

Tethered cord syndrome has been reported in association with neurenteric cysts.[3,9,7,11,15,19] Spinal cord detethering is indicated in patients with progressive neurological deficits[13] and has been shown to improve pain and neurological function in the majority of patients.[11,12] The pathophysiology of tethered cord syndrome is believed to be a result of lack of blood flow from progressive traction.[29] Adult onset tethered cord syndrome is thought to differ from pediatric tethered cord syndrome in that there is a lesser degree of tension on the cord[8] and a certain "threshold" must be crossed before a patient becomes symptomatic.[26] Given that the neurenteric cysts are slow growing lesions that cause symptoms through a compressive process,[23] one could postulate that our patient had a previously asymptomatic tethered spinal cord that was pushed over the threshold from the progressive growth of her neurenteric cyst. We elected to detether the spinal cord prior to intramedullary dissection in the hope that this would reduce the risk for neurological deficits during the lesion resection.

CONCLUSIONS

Intramedullary neurenteric cysts are extremely rare spinal cord lesions that are benign in nature. We report a case of an intramedullary neurenteric cyst in the thorocolumbar region with an associated tethered cord. There are 29 cases in the published literature since the advent of MRI and of the cases with a follow-up MRI, the overall recurrence rate was found to be 25%.

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