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A Rare Intramedullary Epidermoid Cyst of the Thoracic Spinal Cord: Case Report and Review of Literature

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Abstract

Epidermoid cyst in the spinal cord is a rare condition. It constitutes of only 0.6-1.1% of all spinal tumors. When they occur, the typical location is in the subdural, extramedullary space of the lumbo-sacral region. We report a forty-two -year-old female who presented with left extremity radicular pain and myelopathy. MRI of the thoracic spine, illustrated a focal fusiform enlargement of the thoracic cord at T4 and T5. Histopathological examination confirmed the diagnosis of epidermoid cyst. The lesion was surgically resected and the pathology revealed we discuss the clinical features, MR imaging characteristics and surgical findings of this rare tumor and review the associated literature.

Introduction.

Epidermoid cysts are rare tumors of spinal cord. Less than 1% of all spinal cord tumors are epidermoid cysts, (27) which are usually located as intradural and extramedullary and are product of developmental problems (23) and sometimes they are iatrogenic. (8) While most of the reported cases are in thoracic and lumbar area, there are few cases concerning thoracic intramedullary epidermoid cyst. Here we report a case of small intramedullary epidermoid cyst in a 40 year-old woman.

Case report.

A forty-two year-old woman was referred to our clinic for left extremity radicular pain and weakness. The patient had no other complains. Physical exam including neurological examination findings of cranial nerves and upper extremities were normal. Hyperactive reflexes and spasticity of the lower extremities were noted. Bilateral Babinski responses were elicited. The muscle strength of the lower extremities was diminished, especially in distal muscles and the weakness was more significant in the left in contrast to the right side. No definite sensory level was found. A sense of position and vibratory sensations were preserved. The anal reflex was normal.

On lumbosacral MRI, a left paracentral L5-S1 disk protrusion was seen; however, it could not justify her symptoms on the right lower extremity. Therefore, MRI of cervical and thoracic

spines were requested. Cervical MRI findings were normal. Thoracic MRI illustrated a focal fusiform enlargement of the thoracic cord at T₄ and T₅. On sagittal images, hypointensive and hyperintensive intramedullary lesion at T₄ level were seen on T₁ and T₂-weighted images, respectively (figure 1A&B - figure 2). The cord above and below this cavity tapered down to a normal diameter and did not show signal abnormality. The lesion did not show enhancement in MRI with gadolinium (figure 1C).

Operative approach.

A laminectomy (T3-T5) was performed and no evidence of dysraphism, such as a dermal sinus, was found. When the dura was opened, the cord appeared distended (FIGURE 3A). Under the operating microscope, the posterior median raphe was obliterated and the midline was identified at the midpoint between the dorsal root entry zones. A 1.5-cm myelotomy centered over the lesion was performed (FIGURE 3B). Immediately beneath the surface was an avascular mass lying completely within the cord. Pia traction sutures were used to open the myelotomy incision and expose the intramedullary tumor. The mass consisted of a white, waxy, and flaky material, with no apparent skin appendages. The tumor was gradually and completely removed and the cord was decompressed. No attempt was made to remove the lining of the cyst since it was firmly attached to the substance of the cord.

The materials were sent to a pathologist and the diagnosis of epidermoid cyst was confirmed. (figure 4) The patient was visited two weeks later and no symptoms in the left side were seen. The MRI of the brain was normal with no sign of myelin degenerative diseases like multiple sclerosis.

Discussion.

Less than 1 % of intraspinal tumors are epidermoid cysts. These benign tumors are very rare with cranial to spinal ratio of 14:1. (1) Pure intramedullary epidermoid cyst are rare and only 32 intramedullary cysts in thoracic cord are reported table 1. Epidermoid cyst is the result of incomplete cleavage of the neural ectoderm from cutaneous ectoderm from the 3rd to 5th week of gestation. (4) It is usually a developmental problem but iatrogenic cases due to implanting epidermal cells by repetitive lumbar puncture are also reported. (8) MRI is the diagnostic measure of choice to detect the cyst with absence of peri-tumor edema and sharp borders differentiating tumor from normal tissue. (36) The treatment of choice of epidermoid cyst is complete removal, however there is a reports of several recurrence in the same location. (16) While these tumors are essentially considered benign, malignant transformation might be seen. (31)

In our patient, the small epidermoid cyst at T4 was the culprit causing symptoms. The epidermoid cyst may be accompanied by a dermal lesion, which was absent in our case. After the surgery, the patient became symptom free as the pressure on the cord was removed. In 18 month followup, no symptom was seen.

In patients with symptoms and signs of upper and lower motor neuron, before labelling patient with a disease by just brain MRI, a careful imaging study of the spinal cord might be reasonable. In other words, lesion in thoracic region of the spinal cord are usually neglected because of another lesion in the lumbosacral imaging that might justify the symptoms. As in our case, a small intramedullary lesion can mimic sign and symptoms of more serious diseases like multiple sclerosis. The small size of tumor in our patient and normal skin over the thoracic cord in concert with few neurologic symptoms which were to some degree justified by patients' brain MRI had led to misdiagnosis of the condition.

Table1 Summary of the published cases of Epidermoid cyst

	Sex/age	location	Clinical presentation	Cord adhesion	Time to diagnosis	recurrence
Munshi, Anusheel 2018 (26)	Male/3 y	T1-T10	Sudden onset paraparesia	yes	acute	no
Manoj Kumar2018 (23)	Female/22y	T7-T8	Paraparesia&lower limb paresthesia	no	3 Years	no
Atikur Rahman2018 (28)	Male/21y	T2-T3	Paraparesia&bladder dysfunction	yes	6 Mon	no
GRAILLON T2017 (18)	Male/61y	T3-T4	Progressive paraparesia	yes	2y	no
Mohd Faheem2016(12)	Male/27y	T2-T3	Progressive paraparesia	yes	1 Y	no
Rasim Babayev2015 (3)	Female/14 y	T2-T3	Progressive paraparesia	no	2 week	no
Sudhansu Sekhar Mishra2015 (25)	Female/14y	T5	Paresthesia&paresia	yes	4 mon	no
Michael G. Fazio,2013 (13)	Female/32 mon	T2-t8	Recurrent Fever&upper respiratory tract infection	yes	-	yes
Nader-Afshar Fereydoonian2013(14)	Male/40 y	T4	paresthesia	no	2 y	no
Aravind Somas undaram2013 (31)	Male/53 y	T3-T4	paraparesia	no	-	yes
Shams Raza Brohi2010(3)	Male/33 y	T5-T6	Paraparesia&urinaryincontinence	no	12 Y	no
Gonzalvo2009 (17)	Male/40 y	C7-T2	Back pain &lower limb paresthesia	yes	2 Y	no
C.-P. Yen1 2008 (35)	Female/16 y	T3-t4	incontinence and progressive paraparesis.(meningocel surgery)	no	acute	no
MN Swamy2008 (33)	Female/17 y	T4	progressive spastic paraparesis	no	5 y	no
Jun Ho Lee 2008 (22)	Male/53 y	T11-l1	Paraparesia&bladder dysfunction	no	8 mon	no
Rafael Cincu 2006 (10)	Male/24 y	T5-T6	Lower limb paresthesia&progressivepare sia pain in the right thigh and frequent incontinence of urine	No	1 y	no
	Female/28 y	T12-l1		no	6 y	no
ALFRED T. OGDEN2007 (27)	Male/23 y	C7-T1	bowel incontinence´ hand weakness andparesthesiaparesthesia	yes	4 y	no
MING-TSUNG WANG2005 (34)	Female/74y	T11	Progressive paraparesia	no	3 mon	no
Marco Antônio Rocha (19)	Female/15 y	T3-t4	paraparesia	no	8 mon	no

P Ferrara 2003 (15)	Female/13 y	T10-T11	recurrent low urinary tract infections (UTI), urinary frequency and nocturnal enuresis	no	1Y	no
Amato VG2002 (2)	Female/21 y	T3-t4	Paraparesia&paresthesia	yes		no
Alan M. Scarrow 2001 (29)	Female/31y	T4-T5	Progressive paraparesia	no	5 mon	no
Chandra PS 2000 (9)	Female/18 y	T4-T5	progressive difficulty in walking	yes	2 y	no
Zafer Berkman,1997 (5)	Male/40 y	T1-T3	Paraparesia&paresthesia	no	1y	no
Elazhari A 1996 (11)	Male/14 y	T5-T6	Progressive paraparesia	no	6mon	no
J.E Martinez-Lage 1993 (24)	Male/18 mon	C7-t3	difficulty and pain whilst walking +dermal sinus tract	no	3 mon	no
Scholz, M1994 (30)	Femal/32 y	T2-T4	pain in the left knee and difficulty in walking	no	3 mon	no
Buge A 19885 (7)	Male/14 y		Progressive paraparesia	no	2 mon	no
WARD W. STEVENS1968 (32)	Male/40 y	T4-T5	pain and numbness&infrequent bowel incontinence	Yes	18 mon	yes
	Male/40 y	T5-T6	progressive unsteadinessof gait and a more recent onset ourinaryincontinence	yes	5 Y	no
WOLFF M. KIRSCH1965 (20)	Male/6 y	T7	urinary incontinence and increasing weakness ofhis right leg	yes	8mon	no

REVIEW OF THE LITERATURE:

Review of the literature was performed using PubMed by searching for the following combinations: “Epidermoid cyst” AND “spinal cord”. The search was limited to humans and articles reported in English. No restriction was set regarding the type of publication, the publication date, or publication status. The search was broadened by extensively cross-checking the reference lists of all retrieved articles fulfilling the aim of the study. For all databases, the last search was run on 2018.

32 cases of reports described from 1965 to 2018. Table 1 lists all cases reported in the literature with “Epidermoid cyst” AND “spinal cord”. The patient range from 16 months to 74 years old. One

patient presented with fever and recurrent upper respiratory tract infection (13) two patients presented with sudden paresia (17,26) 28 cases of 32 progressive paraparesia was observed. Back pain was observed in 7 cases (17,15,21,22,25,28,35)

In two cases clinical symptoms occurred less than 3 years old. (11,13) They were often referred to a clinic with more severe symptoms and abrupt onset symptoms. In 17 cases of 32 (53%) , urinary tract symptoms such as urinary incontinence and bladder disorders were observed. In one patient the only symptom was recurrent urinary tract infection and incontinence (19) All cases of involvement included the thoracic area in the imaging. In 62.5% (20 patients) of cases, the onset of the complications was reported to be at least 6 months. In 2 patients thoracic intramedullary epidermoid cyst were coexist with sinus dermal tract that diagnosed before 3 years old (11,13) whereas in one history of meningocele surgery in thoracic region was reported (32)

In 19 patient total resection of tumor with tumor capsule excision were achieved but in 13 patients the capsule were adherent to spinal cord and partial resection of capsule were done. 3 patients (7,13,31) underwent second surgery for tumor recurrent in two of them subtotal resection of tumor because of capsule adhesion to spinal cord were done (7,13)

One patient malignant transformation of epidermoid cyst was reported (31)

CONCLUSION :

Spinal intramedullary epidermoid cysts are rare tumors without specific clinical presentation. The recognition of epidermoid cysts in the pediatric age group is important. There may be associated congenital anomalies which need to be carefully sought as an aid to diagnosis. Early diagnosis can be done by MRI. Surgical excision is the treatment of choice and the overall outcome of these lesions is good, even if they present with a ruptured cyst, as was the case with our patient.

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Conflicts of interest

There are no conflicts of interest

Figure 1:



Figure1: sagittal magnetic resonance image (MRI) shows an intramedullary mass lesion in the upper thoracic (T3-T4) with cord expansion. The lesion is hypointense in T1-weighted sagittal MRI (A) and hyperintense in T2-weighted sagittal MRI (B). With contrast, the lesion shows no

Figure 2: enhancement (C).



Figure2: axial magnetic resonance image (MRI) show an intramedullary mass lesion in upper thoracic (T3-T4) with cord expansion. the lesion is hypointense in T1-weighted axial MRI (A) hyperintense in T2-weighted axial MRI (B).

Figure3:

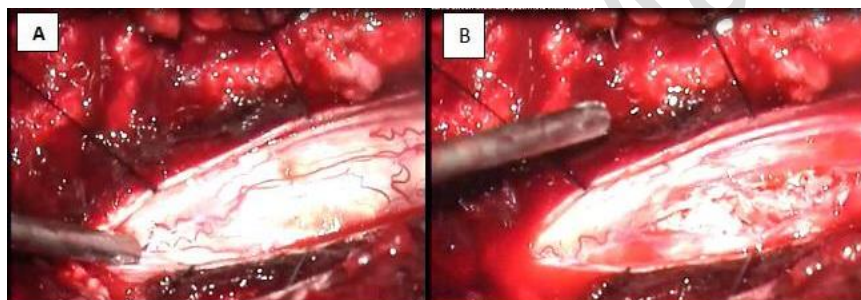
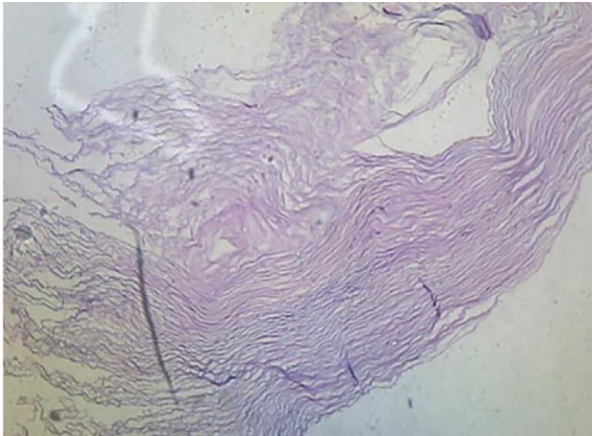


Figure3:A-Intraoperative photograph showing the enlarged spinal cord. B, Intraoperative photograph showing the midline myelotomy, pial traction sutures, and tumor.

Figure4:



The image shows lamellated keratin of intramedullary epidermoid (hematoxylin and eosin stain: ×4)

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