

Case Report:

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



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ABSTRACT

Background and Importance: 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.

Case Presentation: 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.

Conclusion: 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.

Highlights

- Epidermoid cyst can affect the whole neuraxis.
- Epidermoid cyst may be confused with other neurological diseases.
- MRI should be performed in all suspected cases.

Plain Language Summary

Epidermoid cyst of the spinal cord is a rare condition. The typical location of this type of cyst is in the subdural, extramedullary space of the lumbo-sacral region. In this study, the lesion was surgically resected and the pathology was revealed. The clinical features were also discussed.

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1. Background and Importance

Epidermoid cysts are rare tumors of spinal cord. Less than (1%) of all spinal cord tumors are epidermoid cysts [1], which are usually located as intradural and extramedullary and are products of developmental problems [2]. Sometimes they are iatrogenic [3]. As most of the reported cases are in thoracic and lumbar areas, there are few cases concerning thoracic intramedullary epidermoid cysts. Here, we report a case of small intramedullary epidermoid cyst in a 40 year-old woman.

2. Case Presentation

A forty-two year-old woman was referred to our clinic for radicular pain and weakness in left extremity. The patient had no other complaints. Physical examinations including neurological examination 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 in the left in contrast to the right side of the body. 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 the symptoms on her 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 lesions at T4 level were seen on T1 and T2-weighted images, respectively (Figure 1 A & B, Figure 2). The cord above and below this cavity tapered down to a normal diameter and did not show abnormal signal. The lesion did not show any enhancement in MRI with gadolinium (Figure 1 C).

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 3 A). 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 le-

sion was performed (Figure 3 B). There was an avascular mass lying completely within the cord immediately beneath the surface. 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 material was 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 of the body were seen. The MRI of the brain was normal with no sign of myelin degenerative diseases like multiple sclerosis.

3. Discussion

Less than (1%) of intraspinal tumors are epidermoid cysts. These benign tumors are very rare with cranial to spinal ratio of 14:1 [4]. Pure intramedullary epidermoid cysts are rare and only 32 cases of intramedullary cysts in thoracic cord are reported (Table 1). Epidermoid cyst is the result of incomplete cleavage of the neural ectoderm from cutaneous ectoderm within the third to fifth week of gestation [5]. It is usually a developmental problem but iatrogenic cases due to implanting epidermal cells by repetitive lumbar puncture are also reported [3]. MRI is the diagnostic measure to detect the cyst with absence of peri-tumor edema and sharp borders differentiating tumor from normal tissue [6]. The preferred treatment of epidermoid cyst is complete removal. However there is a report of several recurrences in the same location [7]. Although these tumors are essentially considered benign, malignant transformation might be seen [8].

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 surgery, the patient was symptom free as the pressure on the cord was removed. In an 18-month follow-up, no symptom was seen.

In patients with symptoms and signs of upper and lower motor neuron, before labelling the patient with a disease only according to brain MRI findings, a careful imaging study of the spinal cord might be reasonable. In other words, lesions 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 the signs 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 patient's brain MRI had led to misdiagnosis of the condition.

Review of the Literature

Review of the literature was performed using PubMed database 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 extensively cross-checking the reference lists of all retrieved articles fulfilling the aim of the study. For all databases, the latest date of search was run on 2018.

Thirty-two cases of reports belonged to years of publication from 1965 to 2018. Table 1 lists all of the cases reported in the literature with epidermoid cyst and "spinal cord". The patient's age ranged from 16 months to 74 years old. One patient presented with fever and recurrent upper respiratory tract infection [16]. Two patients presented with sudden paresia [9, 19]. Twenty-eight cases of

Table 1. Summary of the published cases of epidermoid cyst

Cases	Sex/ Age	Location	Clinical Presentation	Cord Adhesion	Time to Diagnosis	Recurrence
Munshi, Anusheel [26]	Male/3 y	T1-T10	Sudden onset paraparesia	yes	acute	no
Manoj Kumar [23]	Female/22 y	T7-T8	Paraparesia & lower limb pares- thesia	no	3 y	no
Atikur Rahman [28]	Male/21 y	T2-T3	Paraparesia & bladder dysfunction	yes	6 Mon	no
Graillon [18]	Male/61 y	T3-T4	Progressive paraparesia	yes	2 y	no
Mohd Faheem [12]	Male/27 y	T2-T3	Progressive paraparesia	yes	1 y	no
Rasim Babayev [3]	Female/14 y	T2-T3	Progressive paraparesia	no	2 week	no
Sudhansu Sekhar Mishra [25]	Female/14y	T5	Paresthesia & paresia	yes	4 mon	no
Michael G. Fazio [13]	Female/32 mon	T2-T8	Recurrent fever & upper respiratory tract infection	yes	-	yes
Nader-Afshar Fereydoo- nian [14]	Male/40 y	T4	paresthesia	no	2 y	no
Aravind Somas unda- ram [31]	Male/53 y	T3-T4	paraparesia	no	-	yes
Shams Raza Brohi [3]	Male/33 y	T5-T6	Paraparesia & urinary incontinency	no	12 y	no
Gonzalvo [17]	Male/40 y	C7-T2	Back pain & lower limb paresthesia	yes	2 y	no
Yen [35]	Female/16 y	T3-t4	incontinence and progressive paraparesis (meningocel surgery)	no	acute	no
MN Swamy [33]	Female/17 y	T4	progressive spastic paraparesis	no	5 y	no
Jun Ho Lee [22]	Male/53 y	T11-l1	Paraparesia & bladder dysfunction	no	8 mon	no
Rafael Cincu [10]	Male/24 y	T5-T6	Lower limb paresthesia & progres- sive paresia	No	1 y	no
	Female/28 y	T12-l1	pain in the right thigh and frequent incontinence of urine	no	6 y	no
Alfred, Ogden [27]	Male/23 y	C7-T1	bowel incontinence & acute hand weakness and paresthesia	yes	4 y	no
Ming-Tsung Wang [34]	Female/74 y	T11	Progressive paraparesia	no	3 mon	no

Cases	Sex/ Age	Location	Clinical Presentation	Cord Adhesion	Time to Diagnosis	Recurrence
Marco Antônio Rocha [19]	Female/15 y	T3-T4	Paraparesia	no	8 mon	no
Ferrara (2003) [15]	Female/13 y	T10-T11	Recurrent low Urinary Tract Infections (UTI), urinary frequency and nocturnal enuresis	no	1 y	no
Amato (2002) [2]	Female/21 y	T3-T4	Paraparesia&paresthesia	yes		no
Scarrow (2001)[29]	Female/31y	T4-T5	Progressive paraparesia	no	5 mon	no
Chandra (2008) [9]	Female/18 y	T4-T5	Progressive difficulty in walking	yes	2 y	no
Berkman (1999) [5]	Male/40 y	T1-T3	Paraparesia&paresthesia	no	1 y	no
Elazhari (1996) [11]	Male/14 y	T5-T6	Progressive paraparesia	no	6 mon	no
Martinez-Lage [24]	Male/18 mon	C7-T3	Difficulty and pain whilst walking +dermal sinus tract	no	3 mon	no
Scholz (1994) [30]	Femal/32 y	T2-T4	Pain in the left knee and difficulty in walking	no	3 mon	no
Buge (1985) [7]	Male/14 y		Progressive paraparesia	no	2 mon	no
Stevebs (1968) [32]	Male/40 y	T4-T5	Pain and numbness&infrequent bowel incontinence	yes	18 mon	yes
	Male/40 y	T5-T6	Progressive unsteadiness of gait and a more recent onset ourinaryincontinence	yes	5 y	no
Kirsch [20]	Male/6 y	T7	Urinary incontinence and increasing weakness ofhis right leg	yes	8 mon	no



32 progressive paraparesia were observed. Back pain was observed in seven cases [10, 11, 15, 19, 20, 22, 26].

In two cases, clinical symptoms occurred in less than three years [9, 16, 31]. They were often referred to a clinic with more severe symptoms and abrupt onset symptoms. In 17 of 32 cases (53%), urinary tract symptoms such as uri-



Figure 1. Sagittal Magnetic Resonance Image (MRI) shows an intramedullary mass lesion in upper thoracic (T3-T4) with cord expansion. Hyperintense in T1-weighted sagittal MRI

A. Hyperintense in T2-weighted sagittal MRI; B. The lesion did not show contrast enhancement in MRI; C. The lesion did not show contrast enhancement in MRI with gadolinium



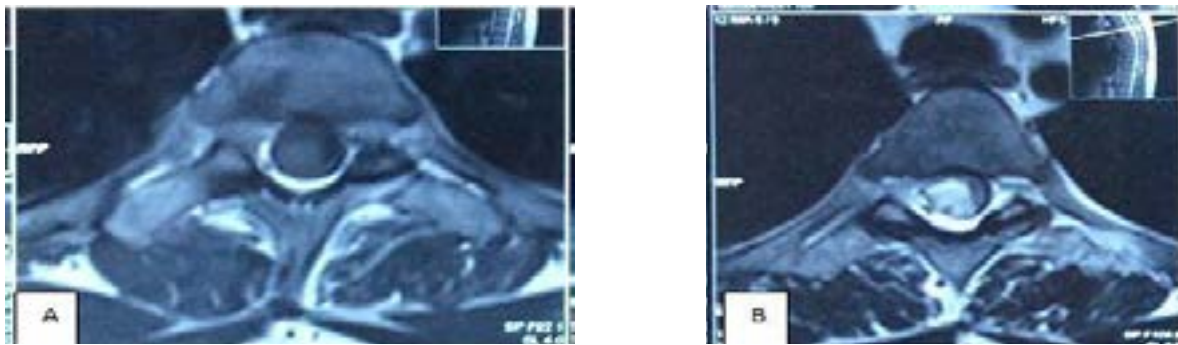


Figure 2. Axial Magnetic Resonance Image (MRI) shows an intramedullary mass lesion in upper thoracic (T3-T4) with cord expansion
A. Hyperintense in T1-weighted axial MRI; B. Hyperintense in T2-weighted axial MRI

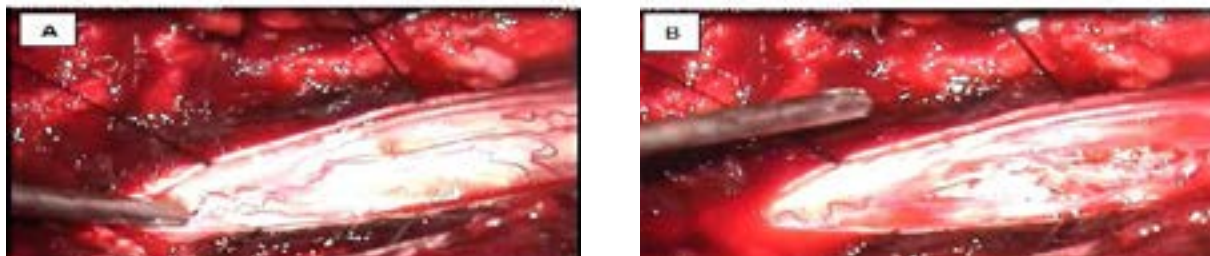


Figure 3. Intraoperative view

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

nary incontinence and bladder disorders were observed. In one patient, the only symptom was recurrent urinary tract infection and incontinency [25]. All cases of involvement included the thoracic area in the imaging. In 20 patients (62.5%) of cases, the onset of the complications was reported to be at least Six months. In two patients, thoracic intramedullary epidermoid cysts coexisted with sinus dermal tract diagnosed before reaching three years old of age whereas in one history of meningocele, surgery in thoracic region was reported [35].

In 19 patients, the total resection of tumor with tumor capsule excision were achieved but in 13 patients the capsules were adherent to spinal cord and partial resection of capsule was performed. Three patients (8, 16, 34) underwent second surgery for tumor recurrent. In two of them, subtotal resection of tumor was done because of capsule adhesion to spinal cord [16, 34]. One patient with malignant transformation of epidermoid cyst was reported [8].

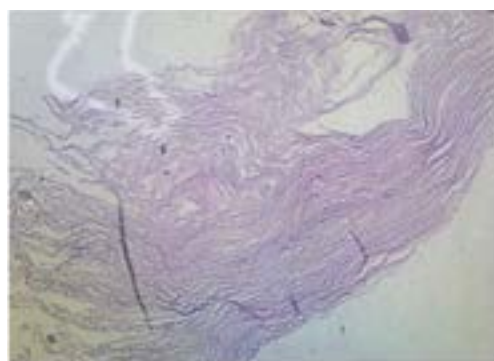


Figure 4. The image showing lamellated keratin of intramedullary epidermoid (hematoxylin and eosin stain: x4)

4. 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.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles were considered in this article. The participants were informed about the purpose of the research and its implementation stages; they were also assured about the confidentiality of their information; Moreover, They were allowed to leave the study whenever they wish, and if desired, the results of the research would be available to them.

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Authors contributions

All authors contributed in preparing this article.

Conflict of interest

The authors declared no conflict of interest.

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