

## 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

**E**pidermoid 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 area, 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 exam 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 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 lesion 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 reports 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 32 progressive paraparesia were observed. Back pain was observed in seven cases [10, 11, 15, 19, 20, 22, 26].

**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 paresthesia	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 Fereydoonian [14]	Male/40 y	T4	paresthesia	no	2 y	no
Aravind Somasundaram [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 & progressive 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
Stevens (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 ourinary incontinence	yes	5 y	no
Kirsch [20]	Male/6 y	T7	Urinary incontinence and increasing weakness of his right leg	yes	8 mon	no



In two, cases clinical symptoms occurred in less than three years old [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

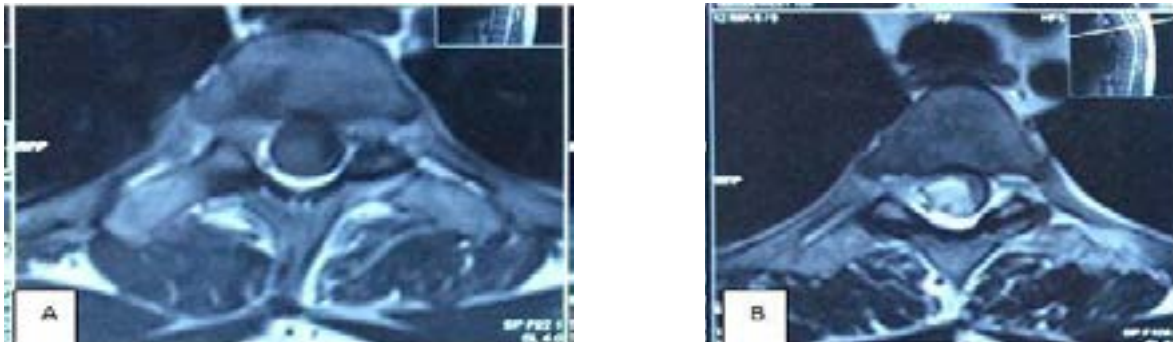
such as urinary incontinence and bladder disorders were observed. In one patient, the only symptom was recurrent urinary tract infection and incontinency [25]. All cases of involvement included the thoracic area in the imaging. In 20



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

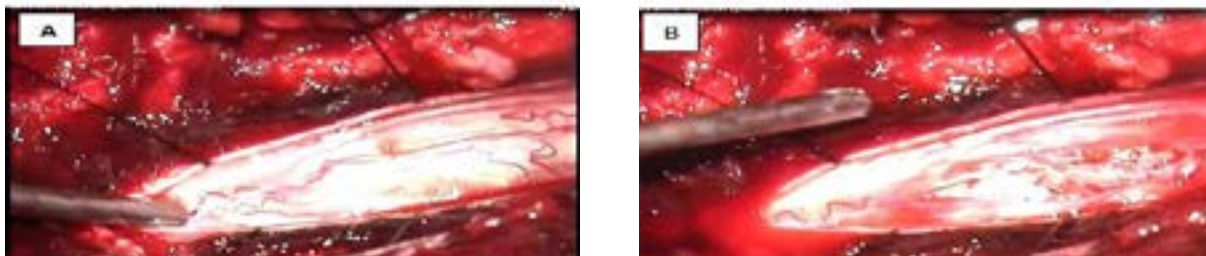
A. Hyperintense in T2-weighted sagittal MRI; B. the lesion did not show contrast enhancement in MRI; C. The lesion did not show contrast enhancement in MRI with gadolinium





**Figure 2.** Axial Magnetic Resonance Image (MRI) shows an intramedullary mass lesion in upper thoracic (T3-T4) with cord expansion. the lesion is

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.

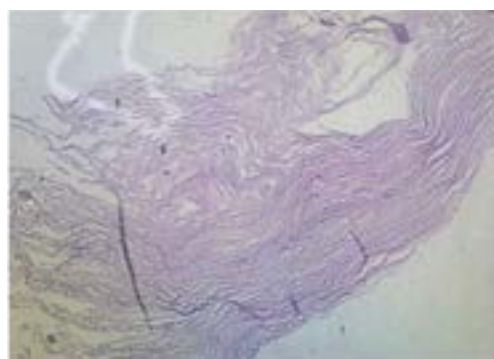
patients (62.5%) of cases, the onset of the complications was reported to be at least Six months. In two patients, thoracic intramedullary epidermoid cysts coexisted with sinus dermal tract diagnosed before reaching three years old of age whereas in one history of meningocele, surgery in thoracic region 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 done. 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 malignant transformation of epidermoid cyst was reported [8].

## 6. Conclusion

Spinal intramedullary epidermoid cysts are rare tumors without specific clinical presentation. The recogni-



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



tion 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.

## References

- [1] Rahman MA, Chaurasia BK, Hossain MA, Habib S, Barua KK. Very rare upperdorsal intramedullary epidermoid with paraplegia: A case report. *Neuroimmunology and Neuroinflammation*. 5, 14. [DOI:10.20517/2347-8659.2017.57]
- [2] Amato VG, Assietti R, Arienta C. Intramedullary epidermoid cyst: Preoperative diagnosis and surgical management after MRI introduction: Case report and updating of the literature/comments. *Journal of Neurosurgical Sciences*. 2002; 46(3/4):122.
- [3] Babayev R, Abbasov B, Ekşi MŞ. Thoracic intramedullary epidermoid cyst-timely fashion diagnosis and treatment. *Child's Nervous System*. 2015; 31(5):793-6. [DOI:10.1007/s00381-015-2625-6] [PMID]
- [4] Berger MS, Wilson CB. Epidermoid cysts of the posterior fossa. *Journal of Neurosurgery*. 1985; 62(2):214-9. [DOI:10.3171/jns.1985.62.2.0214] [PMID]
- [5] Berkman Z, İplikçioğlu C, Bek Ş, Özcan D. Intramedullary epidermoid tumors: A report of two cases. *Marmara Medical Journal*. 1999; 12(1):39-42.
- [6] Buge A, Chamouard JM, Schadeck B, Sichez JP, Fabiani JM. 1985. [Spinal cord congenital epidermoid cyst (a dorsal case)]. *Rev Neurol (Paris)*. 1985; 141(12):810-3. [PMID]
- [7] Çataltepe O, Berker M, Akalan N. A giant intramedullary spinal epidermoid cyst of the cervicothoracic region. *Pediatric Neurosurgery*. 2004; 40(3):120-3. [DOI:10.1159/000079853] [PMID]
- [8] Chandra PS, Manjari T, Devi BI, Chandramouli BA, Srikanth SG, Shankar SK. Intramedullary spinal epidermoid cyst. *Neurology India*. 2000; 48(1):75.
- [9] Cincu R, Lazaro JF, Liesa JL, Callizo JR. Dorsal intramedullary spinal epidermoid cysts: Report of two cases and review of literature. *Indian journal of Orthopaedics*. 2007; 41(4):395. [DOI:10.4103/0019-5413.37005] [PMID] [PMCID]
- [10] Elazhari A, Sami A, Naja A, Ouboukhlik A, Elkamar A, Boucetta M. A thoracic medullary epidermoid cyst. A case report with MRI and literature review. *Neurochirurgie*. 1996; 42(6):309-11.
- [11] Faheem M, Zeeshan Q, Ojha BK, Chandra A, Singh SK, Srivastava C, et al. Isolated thoracic intramedullary epidermoid cyst-a rare entity. *Romanian Neurosurgery*. 2016; 30(2):272-3. [DOI:10.1515/romneu-2016-0042]
- [12] Fazio MG, Kil AM, Rooks VJ, Bięga TJ. Diffusion-weighted magnetic resonance imaging of an intramedullary epidermoid cyst with dorsal dermal sinus tract in a toddler. *Case Rep Radiol*, 2013; 878713. [DOI:10.1155/2013/878713] [PMID] [PMCID]
- [13] Fereydoonian NA, Bakhti S, Fereshtehnejad SM, Tabibkhouei AR. Intramedullary thoracic spine epidermoid cyst with myelopathic presentations: A report of a rare case. *Clinical Neurology and Neurosurgery*. 2013; 6(115):841-3. [DOI:10.1016/j.clineuro.2012.08.002] [PMID]
- [14] Ferrara P, Costa S, Rigante D, Mule A, D'Aleo C, Pulitano S, Belli P, Menchinelli P, Caldarelli M. Intramedullary epidermoid cyst presenting with abnormal urological manifestations. *Spinal Cord*. 2003; 41(11):645. [DOI:10.1038/sj.sc.3101482] [PMID]
- [15] Fleming C, Kaliaperumal C, O'Sullivan M. Recurrent intramedullary epidermoid cyst of conus medullaris. *BMJ Case Reports*. 2011; 2011:bcr1120115090. [DOI:10.1136/bcr.11.2011.5090] [PMID] [PMCID]
- [16] Gonzalvo A, Hall N, McMahon JH, Fabinyi GC. Intramedullary spinal epidermoid cyst of the upper thoracic region. *Journal of Clinical Neuroscience*. 2009; 16(1):142-4. [DOI:10.1016/j.jocn.2008.04.017] [PMID]
- [17] Graillon T, Rakotozanany P, Meyer M, Dufour H, Fuentes S. Intramedullary epidermoid cysts in adults: Case report and updated literature review. *Neurochirurgie*. 2017; 63(2):99-102. [DOI:10.1016/j.neuchi.2017.01.004] [PMID]
- [18] Júnior MA, de Carvalho GT, Faria MP, Faglioni Jr W, de Souza AA. Tumor epidermóide intramedular: relato de caso. *Arquivo Brasileiro De Neuropsiquiatria*. 2003; (61):867-9.
- [19] Kirsch WM, Hodges FJ. An intramedullary epidermal inclusion cyst of the thoracic cord associated with a previously repaired meningocele: case report. *Journal of neurosurgery*. 1966; 24(6):1018-20. [DOI:10.3171/jns.1966.24.6.1018] [PMID]

- [20] Kumar M, Malgonde MS, Kumar SA. Thoracic intramedullary spinal epidermoid tumor: A rare entity. *Saudi Journal for Health Sciences*. 2013; 2, 132. [DOI:10.4103/2278-0521.117920]
- [21] Lee JH, Shin JJ, Hwang YS, Kim TH, Shin HS, Park SK. Intramedullary Epidermoid Cyst in Thoracolumbar Junction: A Case Report. *Korean J Spine*. 2008; 5(3):219-24.
- [22] Manno NJ, Uihlein A, Kernohan JW. Intraspinous epidermoids. *Journal of Neurosurgery*. 1962; 19(9):754-65. [DOI:10.3171/jns.1962.19.9.0754] [PMID]
- [23] Martínez-Lage JF, Esteban JA, Poza M, Casas C. Congenital dermal sinus associated with an abscessed intramedullary epidermoid cyst in a child: Case report and review of the literature. *Child's Nervous System*. 1995; 11(5):301-5.
- [24] Mishra SS, Satapathy MC, Deo RC, Tripathy SR, Senapati SB. Isolated thoracic (D5) intramedullary epidermoid cyst without spinal dysraphism: A rare case report. *Journal of Pediatric Neurosciences*. 2015; 10(2):133.
- [25] Munshi A, Talapatra K, Ramadwar M, Jalali R. Spinal epidermoid cyst with sudden onset of paraplegia. *Journal of Cancer Research and Therapeutics*. 2009; 5(4):290.[
- [26] Ogden AT, Khandji AG, McCormick PC, Kaiser MG. Intramedullary inclusion cysts of the cervicothoracic junction: report of two cases in adults and review of the literature. *Journal of Neurosurgery: Spine*. 2007; 7(2):236-42. [DOI:10.3171/SPI-07/08/236] [PMID]
- [27] Alves AM, Norrell H. Intramedullary epidermoid tumors of the spinal cord. Report of a case and review of the literature. *International surgery*. 1970; 54(4):239. [PMID]
- [28] Scarrow AM, Levy EI, Gerszten PC, Kulich SM, Chu CT, Welch WC. Epidermoid cyst of the thoracic spine: case history. *Clinical Neurology and Neurosurgery*. 2001; 103(4):220-2. [DOI:10.1016/S0303-8467(01)00156-1]
- [29] Scholz M, Märzheuser-Brands S, Gottschalk J, Böck JC, Lanksch WR. Intramedullary epidermoid cyst. A case report. *Neurosurgical Review*. 1994; 17(1):89-93. [DOI:10.1007/BF00309994] [PMID]
- [30] Somasundaram A, Lesser GJ, Mott RT, Hsu W. Malignant transformation of an intramedullary epidermoid cyst in the thoracic region of the spinal cord: Case report. *Journal of Neurosurgery: Spine*. 2013; 19(5):591-4. [DOI:10.3171/2013.8.SPINE13150] [PMID]
- [31] Stevens WW, Schlesinger EB. Intramedullary epidermoid tumors of the thoracic spinal cord: Report of two cases. *Journal of Neurosurgery*. 1968; 29(3):296-9. [DOI:10.3171/jns.1968.29.3.0296] [PMID]
- [32] Swamy MN. A Case of intramedullary epidermoid cyst. *Medical Journal, Armed Forces India*. 2008; 64(1):72.[DOI:10.1016/S0377-1237(08)80155-4]
- [33] Wang M, Wu T, Chen J, Tsai T, Chen T, Tzeng W, Chang J, Su C. Magnetic resonance imaging of intramedullary epidermoid cyst of low thoracic spinal cord: a case report. *Chinese Journal of Radiology-Taipei*. 2005; 30(2):109.
- [34] Yen CP, Kung SS, Kwan AL, Howng SL, Wang CJ. Epidermoid cysts associated with thoracic meningocele. *Acta neurochirurgica*. 2008; 150(3):305-9. [DOI:10.1007/s00701-007-1398-4] [PMID]
- [35] Zavanone M, Guerra P, Rampini PM, Crotti F, Vaccari U. A cervico-dorsal intramedullary epidermoid cyst. Case report and review of the literature. *Journal of Neurosurgical Sciences*. 1991; 35(2):111-5.

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