



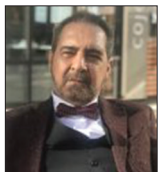
Review Article

# Spinal intramedullary epidermoid cysts: Three case presentations and literature review

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## ABSTRACT

**Background:** True intramedullary epidermoid cysts (IECs) not associated with congenital anomalies or previous spinal procedures are extremely rare. In a review of the literature since 1992, only 29 such cases have been reported. Here, we add three new cases in this category.

**Case Description:** Three adults presented with spastic paraparesis attributed to thoracic IECs. Gross total microsurgical removal was achieved in two cases, while one case was a partial resection due to capsular adherence to the cord. In all three cases, patients sustained complete recoveries of neurological function and remained symptom free for an average of 5 years follow-up.

**Conclusion:** IECs are rare lesions; here, the three located in the thoracic spine, contributed to slow, progressive spastic paraparesis with/without incontinence, and resolved following total (2 patients) and partial (1 patient) resection.

**Keywords:** Epidermoid cyst, Intramedullary tumor, Paraparesis, Spinal cord, Thoracic spine

## INTRODUCTION

Spinal epidermoid tumors are rare benign lesions, representing < 1% of all intraspinal tumors; most are intradural and extramedullary in location.<sup>[16,24,34,36]</sup> However, true intramedullary epidermoid cysts (IECs) occurring without spinal dysraphism or prior surgery are even more infrequent, comprising 0.8% of all spinal epidermoid tumors.<sup>[4,7,8,13,16,17,21,22,24,33-36]</sup>

## CASE DESCRIPTION

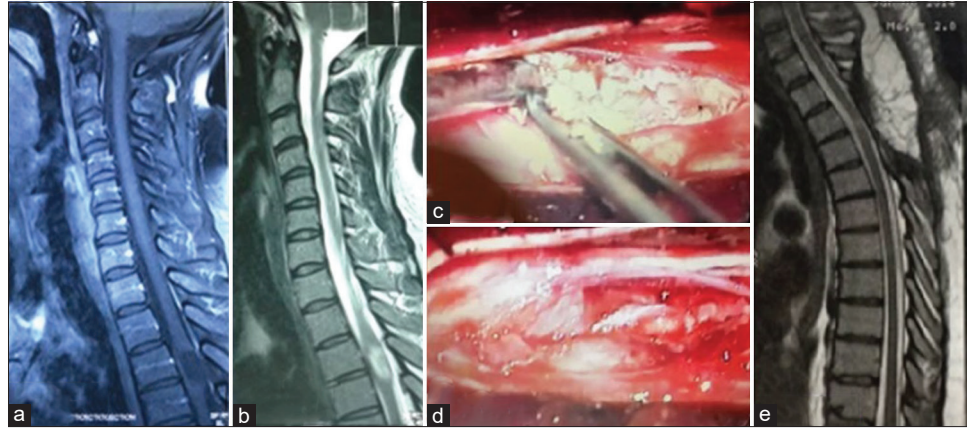
Here, we present three cases of true IEC and review the literature concerning their overall clinical/radiographic presentation and surgical management.

### Case 1

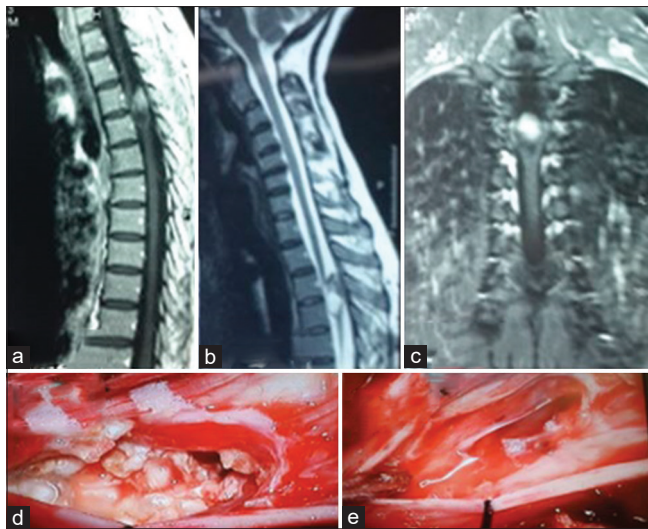
A 40-year-old female presented with a progressive spastic paraparesis and a sensory level bilaterally at T5 over 1 year (e.g. American Spinal Injury Association [ASIA] D classification). The magnetic resonance imaging (MRI) revealed a well-circumscribed intramedullary tumor

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**Figure 1:** (a) Cervicothoracic T1-weighted magnetic resonance imaging (MRI) a hypointense tumor at T3-T4 level, (b) in T2-weighted MRI the mass is hyperintense, (c) intraoperative photograph; a white avascular tumor inside the cord, (d) after total removal, (e) postoperative T2-weighted MRI which is clear from residue.



**Figure 2:** (a) Cervicothoracic T1-weighted sagittal magnetic resonance imaging (MRI) shows a mixed hypo-hyperintense mass at T3-T4 level, (b) in T2-weighted image, the mass is also hypo-hyperintense which is not usual in intramedullary epidermoid cysts, (c) but in fat-suppressed MRI the whole of the tumor is hyperintense, (d) intraoperative photograph; a milky white tumor is demonstrated after midline myelotomy, (e) after total removal of the mass.

at the T3-T4 level that was hypointense on T1 weighted and hyperintense on T2-weighted sequences [Figure 1a and b]. After T3 and T4 laminectomy and dural opening, the cord was enlarged and incised longitudinally in the midline allowing for the identification of a white cheese-like avascular tumor [Figure 1c]. Utilizing a microscope, piecemeal removal was achieved [Figure 1d], leaving a few small patches of capsule densely adherent to the cord, behind. The postoperative course was uneventful, and she regained full function within 3 months that was maintained

at 2 postoperative years. Further, the postoperative MRI 1 week later shows no residual tumor [Figure 1e].

### Case 2

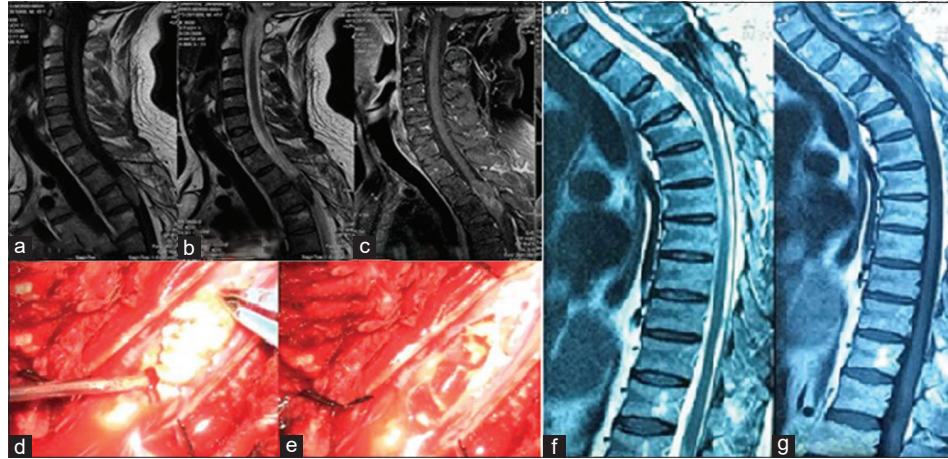
Progressively over a 3-year period, a 37-year-old female also presented with a spastic paraparesis and paresthesia/sensory level T5 bilaterally (ASIA C classification). The cervicothoracic sagittal MRI showed an intramedullary mass at the T3-T4 level; there was a mixture of hypo and hyperintensities on the T1- and T2-weighted images [Figure 2a and b]. On the fat-suppressed MRI, the entire mass was hyperintense [Figure 2c]. She too underwent T3 and T4 laminectomy with midline myelotomy; an avascular white tumor with sebaceous consistency was found and completely removed, including the entire capsule [Figure 2d and e]. The patient gradually improved over the next 6 months; she was able to ambulate without support (ASIA Class D). Five years later, she was intact (ASIA Class E).

### Case 3

A 41-year-old male who also was paraparetic, had an intramedullary IEC at the T3-T4 level that was similarly grossly totally removed, resulting in the patient's eventual full and sustained recovery at 8 years follow-up (ASIA Class E) [Figure 3].

## DISCUSSION

In 1992, Roux *et al.* reviewed all true IECs within medical literature and could find 47 cases including a case of their own.<sup>[34]</sup> We updated the literature and could add 31 more cases including our three new ones [Table 1].<sup>[16,24,36]</sup> Detailed information about the gender, age, and location of the tumor is shown in separately [Table 2].



**Figure 3:** (a) Cervicothoracic T1-weighted magnetic resonance imaging (MRI) the tumor is isointense, (b) in T2-weighted mage it is hypointense which is unusual for intramedullary epidermoid cysts, (c) but in fat-suppressed MRI the hyperintense mass is compatible with epidermoid, (d) intraoperative photograph shows the characteristic features of and epidermoid cyst, (e) after total removal, (f) T1-weighted MRI at 10-year follow-up shows neither residue nor recurrence, (g) T2-weighted image also is clear.

**Table 1:** Review of the literature: Published cases.

Author	Year	Sex	Age	Location	Clinical picture	Management	Outcome
Scholz <i>et al.</i> <sup>[35]</sup>	1994	M	32	T3-T4	Paraparesis	Laminectomy	Good
Jadhav <i>et al.</i> <sup>[17]</sup>	1999	F	12	C7-T2	Quadriparesis	Laminectomy	Good
Chandra <i>et al.</i> <sup>[8]</sup>	2000	F	18	T4-T5	Paraparesis	Laminectomy	Good
Chandra <i>et al.</i> <sup>[8]</sup>	2000	F	28	Conus	Paraparesis and incontinence	Laminectomy	Good
Amato <i>et al.</i> <sup>[4]</sup>	2002	F	21	T3-T4	Paraparesis	Laminectomy	Good
Rocha <i>et al.</i> <sup>[33]</sup>	2003	F	15	T2-T4	Monoparesis	Laminectomy	Good
Ferrara <i>et al.</i> <sup>[13]</sup>	2003	F	13	T10-T11	Incontinence	Laminectomy	Fair
Cataltepe <i>et al.</i> <sup>[7]</sup>	2004	M	6	C1-T5	Quadriparesis	Laminoplasty	Good
Kumar and Sing <sup>[21]</sup>	2004	M	4	Conus	Paraparesis and incontinence	NM	Good
Kumar and Sing <sup>[21]</sup>	2004	F	14	T3-T5	Paraparesis	NM	Good
Lai <i>et al.</i> <sup>[22]</sup>	2005	M	49	Conus	Monoparesis+incontinence	Laminectomy	Fair
Moon <i>et al.</i> <sup>[26]</sup>	2006	F	43	Conus	Incontinence	Laminectomy	Poor
Cincu <i>et al.</i> <sup>[9]</sup>	2007	M	27	T5-T6	Monoparesis	Laminectomy	Good
Ogden <i>et al.</i> <sup>[28]</sup>	2007	F	61	C3-T1	Paraparesis	Laminectomy	Fair
Lee <i>et al.</i> <sup>[23]</sup>	2008	M	53	Conus	Paraparesis and incontinence	Laminectomy	Good
Gonzalvo <i>et al.</i> <sup>[14]</sup>	2009	M	40	C7-T2	Monoparesis+incontinence	Laminectomy	Good
Kumar <i>et al.</i> <sup>[20]</sup>	2010	F	10	C6-T5	Paraparesis+meningitis	Laminectomy	Fair
Brohi <i>et al.</i> <sup>[6]</sup>	2010	M	33	T5-T6	Paraplegia	Laminectomy	Poor
Agarwal <i>et al.</i> <sup>[11]</sup>	2011	F	40	C2-C3	Quadriparesis	Laminectomy	Good
Fereydoonian <i>et al.</i> <sup>[12]</sup>	2012	M	40	T4	Paraparesis	Laminectomy	Good
Yoon <i>et al.</i> <sup>[37]</sup>	2013	F	55	C4-T10	Paraplegia	Laminectomy	Good
Gotecha <i>et al.</i> <sup>[15]</sup>	2014	F	23	Conus	Paraparesis+incontinence	Laminectomy	Good
Babayev <sup>[5]</sup>	2015	F	14	T2-T3	Monoparesis	Laminectomy	Good
Ohara <i>et al.</i> <sup>[29]</sup>	2015	F	63	Conus	Paraparesis+incontinence	Laminectomy	Good
Mishra <i>et al.</i> <sup>[25]</sup>	2015	M	14	T4-T5	Paraparesis	Laminectomy	Good
Jain <i>et al.</i> <sup>[18]</sup>	2016	M	22	T2	Mild paraparesis	Laminectomy	Good
Elsebaey and Elgohary <sup>[10]</sup>	2017	M	42	T4-T6	Paraparesis	Laminectomy	Good
Agrawal <i>et al.</i> <sup>[2]</sup>	2019	F	32	Conus	Paraparesis and incontinence	Laminectomy	Poor
Musali <i>et al.</i> <sup>[27]</sup>	2019	F	6	Conus	Paraparesis	Laminectomy	Good
Current case	2019	F	40	T3-T4	Paraparesis	Laminectomy	Good
Current case	2019	F	37	T3-T4	Paraparesis	Laminectomy	Good
Current case	2019	M	41	T2-T3	Paraparesis	Laminectomy	Good

M: Male, F: Female, NM: Newton meter

**Table 2:** Descriptive statistics.

	Frequency	Percentage
Age		
≤18 years	11	34.4
>18 years	21	65.6
Sex		
Male	13	40.6
Female	19	61.3
Location		
Cervical	1	3.2
Cervicothoracic	6	19.4
Thoracic	16	54.8
Conus	9	22.6
Clinical picture		
Monoparesis	3	9.4
Paraparesis	13	40.6
Quadriparesis	3	9.4
Paraparesis+incontinence	6	18.8
Monoparesis+incontinence	2	6.3
Paraparesis+meningitis	1	3.1
Paraplegia	2	6.3
Incontinence	2	6.3
Outcome		
Fair	4	12.5
Good	25	78.1
Poor	3	9.4
Total	32	100.0

MRI characteristically demonstrates a nonhomogeneous, hypodense, or isointense mass on T1-weighted MRI scans attributed to the variable amounts of the lipids and proteins within the tumor. They are hyperintense on T2-weighted image due to the keratin content of the cyst.<sup>[1,2,4-10,12-15,17,18,20-23,25-29,33,35,37]</sup> The diffusion-weighted MR best demonstrates epidermoid cyst homogeneous hyperintensity.<sup>[11,19]</sup> With these characteristic features, IECs can be easily differentiated from intramedullary lipoma, teratoma, and arachnoid cyst.<sup>[30-32]</sup> ECs usually have relatively sharp boundaries without any edema and a minimal amount of rim enhancement with a gadolinium. Calcification is extremely.<sup>[2]</sup>

### Surgical management

Surgical removal is the optimal management of IECs. At surgery, following midline myelotomy, the tumor is typically well demarcated; with a smooth, hypovascular, and capsule that is readily removed in over half of the patients. The other half may exhibit dense adhesions of the capsule to the cord, precluding total excision.<sup>[3]</sup> During operative dissection, spillage into the subarachnoid space is critical to prevent a postoperative chemical meningitis. Histologically, IECs have a thin capsule of stratified, keratinized, and squamous epithelium that contains an accumulation of desquamated epithelial cells, abundant keratin, small foci of calcifications, and cholesterol clefts.

### CONCLUSION

IECs, most often found in the thoracic spine, typically contribute to progressive paraparesis. MR studies typically demonstrate well-circumscribed lesions that may be readily totally or partially excised, resulting in marked postoperative neurological recovery.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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