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. 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.

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...
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 the 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. We updated the literature and could add 31 more cases including our three new ones [Table 1]. Detailed information about the gender, age, and location of the tumor is shown in separately [Table 2].
Table 1: Review of the literature: Published cases.

| Author               | Year | Sex | Age | Location   | Clinical picture                  | Management     | Outcome |
|----------------------|------|-----|-----|------------|-----------------------------------|----------------|---------|
| Scholz et al.        | 1994 | M   | 32  | T3-T4      | Paraparesis                       | Laminectomy    | Good    |
| JadHAV et al.        | 1999 | F   | 12  | C7-T2      | Quadriparesis                     | Laminectomy    | Good    |
| Chandra et al.       | 2000 | F   | 18  | T4-T5      | Paraparesis                       | Laminectomy    | Good    |
| Chandra et al.       | 2000 | F   | 28  | Conus      | Paraparesis and incontinence      | Laminectomy    | Good    |
| Amato et al.         | 2002 | F   | 21  | T3-T4      | Paraparesis                       | Laminectomy    | Good    |
| Rocha et al.         | 2003 | F   | 15  | T2-T4      | Monoparesis                       | Laminectomy    | Good    |
| Ferraro et al.       | 2003 | F   | 13  | T10-T11    | Incontinence                      | Laminectomy    | Fair    |
| Cataltepe et al.     | 2004 | M   | 6   | C1-T5      | Quadriparesis                     | Laminoplasty   | Good    |
| Kumar and Sing       | 2004 | M   | 4   | Conus      | Paraparesis and incontinence      | NM             | Good    |
| Kumar and Sing       | 2004 | F   | 14  | T3-T5      | Paraparesis                       | NM             | Good    |
| Lai et al.           | 2005 | M   | 49  | Conus      | Monoparesis and incontinence      | Laminectomy    | Fair    |
| Moon et al.          | 2006 | M   | 43  | Conus      | Incontinence                      | Laminectomy    | Poor    |
| Cincu et al.         | 2007 | M   | 27  | T5-T6      | Monoparesis                       | Laminectomy    | Good    |
| Ogden et al.         | 2007 | F   | 61  | C3-T1      | Paraparesis                       | Laminectomy    | Fair    |
| Lee et al.           | 2008 | M   | 53  | Conus      | Paraparesis and incontinence      | Laminectomy    | Good    |
| Gonzalvo et al.      | 2009 | M   | 40  | C7-T2      | Monoparesis and incontinence      | Laminectomy    | Good    |
| Kumar et al.         | 2010 | F   | 10  | C6-T5      | Paraparesis and meningitis        | Laminectomy    | Fair    |
| Brohi et al.         | 2010 | M   | 33  | T5-T6      | Paraplegia                        | Laminectomy    | Poor    |
| Agarwal et al.       | 2011 | F   | 40  | C2-C3      | Quadriparesis                     | Laminectomy    | Good    |
| Fereydoonian et al.   | 2012 | M   | 40  | T4         | Paraparesis                       | Laminectomy    | Good    |
| Yoon et al.          | 2013 | F   | 55  | C4-T10     | Paraplegia                        | Laminectomy    | Good    |
| Gotecha et al.       | 2014 | F   | 23  | Conus      | Paraparesis and incontinence      | Laminectomy    | Good    |
| Babayev              | 2015 | F   | 14  | T2-T3      | Monoparesis                       | Laminectomy    | Good    |
| Obara et al.         | 2015 | F   | 63  | Conus      | Paraparesis and incontinence      | Laminectomy    | Good    |
| Mishra et al.        | 2015 | M   | 14  | T4-T5      | Paraparesis                       | Laminectomy    | Good    |
| Jain et al.          | 2016 | M   | 22  | T2         | Mild paraparesis                  | Laminectomy    | Good    |
| Elsebaey and Elghory  | 2017 | M   | 42  | T4-T6      | Paraparesis                       | Laminectomy    | Good    |
| Agrawal et al.       | 2019 | F   | 32  | Conus      | Paraparesis and incontinence      | Laminectomy    | Poor    |
| Musali et al.        | 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
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. The diffusion-weighted MR best demonstrates epidermoid cyst homogeneous hyperintensity. With these characteristic features, IECs can be easily differentiated from intramedullary lipoma, teratoma, and arachnoid cyst. ECs usually have relatively sharp boundaries without any edema and a minimal amount of rim enhancement with a gadolinium. Calcification is extremely rare.

### 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. 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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### Conflicts of interest

There are no conflicts of interest.

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