

## REVIEW ARTICLE

# Spinal intradural extramedullary mature cystic teratoma in an adult: A rare tumor with review of literature

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## ABSTRACT

**Background:** Spinal intradural extramedullary teratoma is a rare condition, more common in children than in adults often with a history of spinal dysraphism.

**Method:** We reviewed the literature and Pubmed advanced search showed nine results of intradural extramedullary teratoma in adults which included five independent case reports and three cases in a case series. We reported a rare case of intradural extramedullary teratoma in an adult located opposite L4 vertebra with neither spinal dysraphism nor any history of the prior spinal procedure.

**Results:** The occurrence of teratomas in the spine is extremely rare. Further, spinal intradural extramedullary teratoma is more common in children but a rare entity in adults.

**Conclusion:** Although uncommon, spinal cord neoplasm should be considered in the differential diagnosis of backache or radicular pain associated with neurological deficits even in absence of spinal dysraphism or any spinal procedure.

**Key words:** Adults, intradural extramedullary, spinal, teratoma

## Background

The incidence of intracranial teratomas is low, approximately 0.5–2.2% of all intracranial tumors.<sup>[1]</sup> The occurrence of teratomas in the spine is extremely rare.<sup>[2]</sup> Except for in the sacrococcygeal region, teratomas constitute < 0.5% of all intraspinal tumors.<sup>[3]</sup>

## Case Presentation

A 30-year-old adult male presented with complaints of tingling and numbness in left lower extremity since last 6 months which progressed further within next 2 months to involve the right lower extremity. 2 months later, the patient started

experiencing difficulty in walking and standing from sitting position, associated with shooting pain in the lower back region. No bowel and bladder involvement was present. There was no history of any congenital spinal deformity, any spinal surgery or any other spinal procedures like a lumbar puncture.

On physical examinations, there was no motor or sensory deficit in either extremity. No cutaneous abnormalities or dermal sinus tracts were found. No congenital spinal deformity was seen. Neurological examination showed normal superficial and deep tendon reflexes. The laboratory parameters were unremarkable.

The magnetic resonance imaging (MRI) scan revealed a well-defined intradural extramedullary mass lesion measuring 2.7 cm cranio-caudally × 1.6 cm transversely × 1.1 cm anteroposteriorly, located opposite L<sub>4</sub> vertebrae level predominantly on right side leading to compression and displacement of cauda equina nerve roots toward left side. The mass was hypointense to cord on T1-weighted images and hyperintense on T2-weighted images and showed minimal enhancement on postgadolinium images [Figure 1a-f]. There was no lateral extension of the mass into lateral recess. The rest of spinal cord, cord termination, and conus medullaris were normal in signal thus ruling out spinal cord edema or myelomalacia. Intervertebral discs were found to be normal. No prevertebral and paravertebral collection was

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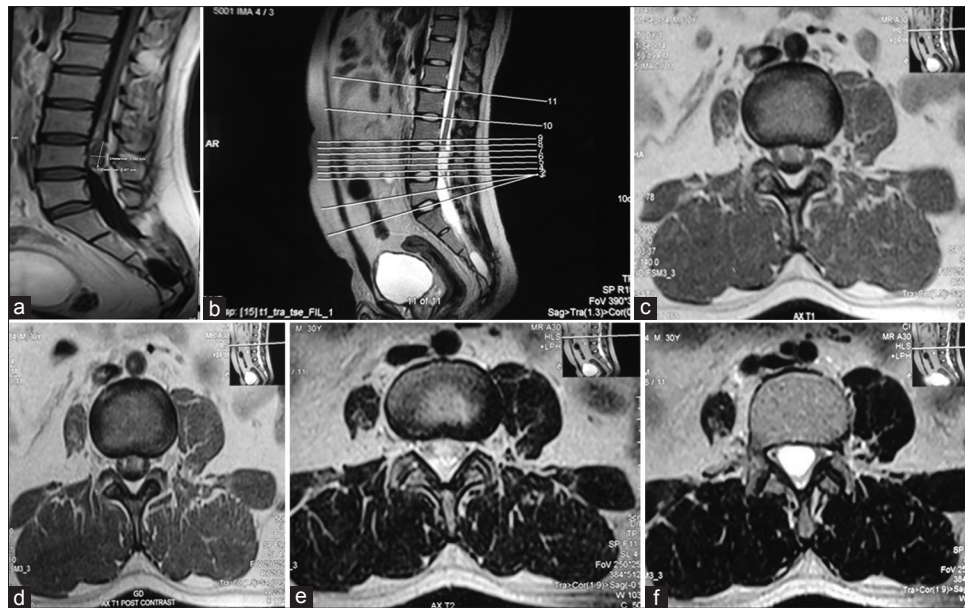
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**Figure 1:** (a) Sagittal T1-weighted magnetic resonance imaging scan revealing a hypointense intradural extramedullary mass located opposite L<sub>4</sub> vertebrae. (b) Sagittal T2-weighted magnetic resonance imaging image revealing a hyperintense well-defined intradural extramedullary mass lesion. (c) Axial T1-weighted magnetic resonance imaging scan revealing a hypointense mass located opposite L<sub>4</sub> vertebrae. (d) Axial T1-weighted magnetic resonance imaging scan (postgadolinium) revealing a hypointense mass located opposite L<sub>4</sub> vertebrae with minimal peripheral enhancement. (e-f) Axial T2-weighted magnetic resonance imaging scan revealing an hyperintense intradural extramedullary mass located opposite L<sub>4</sub> vertebrae

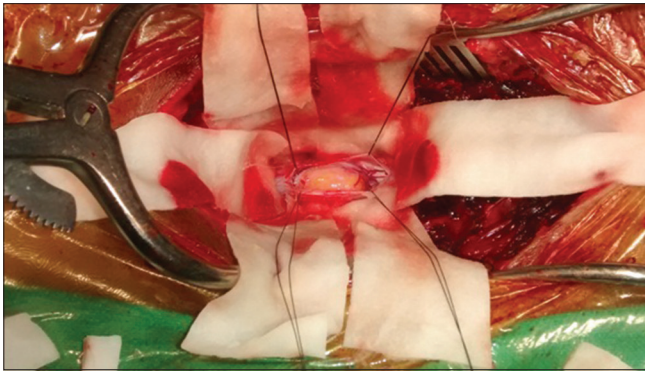
present. Surgical intervention was planned, and the patient underwent total resection of the tumor along with L<sub>3</sub> partial and L<sub>4</sub> complete laminectomy performed in the prone position. On incising the duramater via midline incision, a yellow, oval-shaped, fatty cyst was observed [Figure 2]. Following the incision into the tumor tissue, whitish fluid containing grayish soft tumor tissue was found [Figure 3]. Separation of the capsule and total removal of the tumor were possible. The histopathology showed ciliated pseudostratified columnar epithelium, keratin debris, adipose tissue, and glandular tissue [Figure 4]. Histopathology examination suggested that the overall features were consistent with mature cystic teratoma [Figures 4a-d]. The dura mater was closed in a watertight manner. Postoperative period was uneventful, and the patient was discharged on day 7 without any neurological deficit or signs of meningitis. No further neurological deterioration was observed during the 3-month follow-up period.

## Discussion and Review of Literature

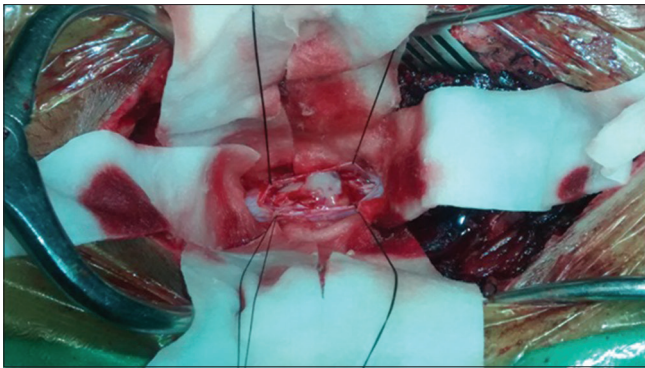
Spinal tumors can be extra-dural or intradural. Intradural tumors can further be classified as intramedullary or extramedullary. Among the wide variety of intradural-extramedullary tumors in adults, the most common are nerve sheath tumors (neurofibromas and schwannomas) and meningiomas<sup>[4]</sup> with spinal teratoma being a rare one. Intradural-extramedullary tumors are the most common primary neoplasm in the spine, accounting for approximately 55% of primary spinal tumors.<sup>[5]</sup>

The first reported case of spinal teratoma was described in 1863 by Virchow.<sup>[6]</sup> Spinal intradural extramedullary teratoma is a rare entity in adults. It is more common in children and often associated with spinal dysraphism.<sup>[7]</sup> The association of teratomas and spinal malformations such as spina bifida, partial sacral agenesis, hemivertebrae, myelomeningocele, tethered cord syndrome, and diastematomyelia has been described in the literature.<sup>[8-12]</sup> Besides congenital abnormality, cases have been reported where a history of trauma or surgical interventions (example lumbar puncture) often preceded the clinical presentation in adults.<sup>[13,14]</sup> Iatrogenic cysts may be caused by the inclusion of some dermal or epidermal tissue during the closure of a myelomeningocele or by the introduction of the epithelium during epidural spinal injections.<sup>[15]</sup> Epidermoid and dermoid cyst that constitute <1% of spinal tumors may be congenital (hamartoma) or iatrogenic in nature.<sup>[4]</sup> The clinical features, including sensory changes depend on the location of the tumors. Our patient presented with tingling and numbness, low backache, progressive weakness in legs. We reported a rare case of spinal intradural extramedullary cystic teratoma in an adult without any spinal dysraphism or history of spinal procedure.

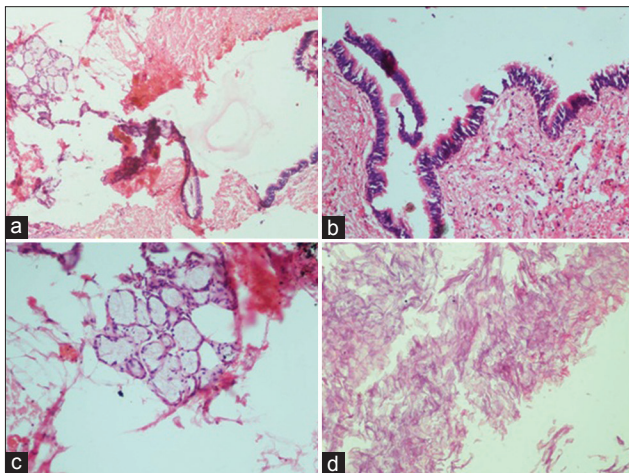
We reviewed the literature and Pubmed advanced search showed nine results of intradural extramedullary teratoma in adults which included five independent case reports<sup>[7,14,16-18]</sup> and three cases in a case series<sup>[19]</sup> [Table 1]. We excluded all cases below 18 years of age. Interestingly, Stevens *et al.* reported the first case of a spinal intradural extramedullary cystic teratoma



**Figure 2:** Intra-operative photograph showing a yellow, oval-shaped, fatty cyst on incising the dura mater



**Figure 3:** Intra-operative photograph showing whitish fluid containing grayish soft tumor tissue following incision into the tumor tissue



**Figure 4:** (a) Microphotograph showing ciliated pseudostratified columnar epithelium and lobules of mucosal epithelial glands embedded in adipose tissue (H and E,  $\times 100$ ). (b) Ciliated pseudostratified columnar epithelium (H and E,  $\times 200$ ). (c) Mucosal glands (H and E,  $\times 200$ ). (d) Keratin debris noted in the other part of the section (H and E,  $\times 100$ )

in an aged patient without any previous lumbar procedures and no history of spinal dysraphism.<sup>[14]</sup>

Li *et al.* reviewed the literature in adult intradural teratoma cases from 1928 to 2013 and found that compared to children, the incidence of mature intraspinal teratomas in adults was

rare, presenting typically with slow onset, more localised lesion, commonly located between lower thoracic and conus medullaris, numbness or weakness of the lower-extremities being the main symptom occasionally accompanied by pain, without an obvious decline of motor grade, rarely observed with vertebral body anomalies or thoracolumbar spinal bifida.<sup>[20]</sup> All these observations were quite consistent with the present case and our review of the literature on adult intradural extramedullary teratoma cases except onset of the disease was slow only in one case.

The differential diagnosis of lumbar radiculopathy in this case includes degenerative conditions like disc herniation, inflammatory conditions like multiple sclerosis, transverse myelitis, infective conditions like an abscess, tuberculosis, vascular conditions like cord infarction and neoplasms.<sup>[21]</sup> Although uncommon, spinal cord neoplasm should be considered the differential diagnosis in patients presenting back or radicular pain associated with neurological deficits.<sup>[22]</sup> In our case, an adult patient, clinical history, physical examination, and laboratory results increased our suspicion of degenerative condition or neoplasms, which was resolved with the help of imaging studies.

The MRI findings of intradural extramedullary tumors include displacement of the cord to the contralateral side of the thecal sac, widening of the space available for the cerebrospinal fluid above and below the tumor, and a sharp demarcation between the tumor and the cerebrospinal fluid. With larger tumors, the spinal cord is often flattened against the dura mater on the contralateral side.<sup>[4]</sup> In our case, MRI scan revealed a well-defined intradural extramedullary mass lesion, located opposite L<sub>4</sub> vertebrae level predominantly on right side which was compressing and displacing cauda equina nerve roots thus accounting for progressive sensory symptoms of the patient. However, bowel and bladder involvement was not found in this patient. MRI is regarded as the gold standard diagnostic technique that can reveal the location of teratomas and consequently the degree of spinal cord involvement.<sup>[7]</sup> Preoperative diagnosis of spinal teratoma is not easy since the MRI features cannot determine with certainty the differential diagnosis between teratoma and other extramedullary lesions.<sup>[6]</sup> The performance of a histopathological examination subsequent to surgery is the final analysis required to confirm the diagnosis of an intraspinal mature teratoma.<sup>[23]</sup>

The diagnosis of teratoma depends on the histopathological identification of the tissues representing the three germinal layers (ectoderm, mesoderm, and endoderm).<sup>[24]</sup> However, the presence of just two layers does not rule out the diagnosis.<sup>[10]</sup> Li *et al.* analyzed the literature and found that in a number of cases, only two of the three germinal layers were observable and attributed this to the fact that the derivatives of one or two of the layers had grown over the others.<sup>[10,20,25]</sup>

**Table 1: Review of intradural extramedullary teratoma cases (18-85 years)**

Year	Author	Number of cases	Age/sex	Onset	Location	Associated abnormality
2006	Kumar <i>et al.</i> <sup>[16]</sup>	01	19/female	Rapid progressive lesion during pregnancy	IDEM lesion at T5, T6	Spinal dysraphism present
2006	Stevens <i>et al.</i> <sup>[14]</sup>	01	85/male	Progressive	IDEM mass at L1, L2	Absent
2008	Sung <i>et al.</i> <sup>[7]</sup>	01	38/male	Adult onset	IDEM mass at T11, T12 and L1	Absent
2009	Liu <i>et al.</i> <sup>[19]</sup>	03	Individual case details not found			
2011	Bouaziz <i>et al.</i> <sup>[17]</sup>	01	38/female	Adult onset	Adult onset IDEM mass cervical spinal cord (on both sides of foramen magnum) compressing bulbomedullary junction	Absent
2014	Vanguardia <i>et al.</i> <sup>[18]</sup>	01	41/male	Slow onset	IDEM at T12/L1 causing marked compression of distal conus	Absent

IDEM – Intradural-extramedullary

The primary treatment modality for symptomatic patients is total surgical resection. In this patient, complete tumor resection was planned with L<sub>3</sub> partial and L<sub>4</sub> complete laminectomy. Intimal adhesion of the teratomas to the surrounding neural parenchyma is observed about 50% cases, thus making it difficult to resect totally.<sup>[8-10,26]</sup> Since, subtotal resection of these tumors increases the chance of recurrence, so they recommend that it is important to resect the tumors as much as possible while preserving all the surrounding neural tissue.<sup>[9,10,14]</sup> During surgery, care should be taken to prevent the cystic contents from spilling into the intradural space to avoid the occurrence of aseptic chemical meningitis with or without obstructive hydrocephalus.<sup>[13,14,24]</sup> In the present case, complete resection was achieved without the injury to adjacent neural tissues, and thus, no further neurological defects were observed following the surgery. The excised tumor tissue was sent for histopathological analysis which revealed the nature of this tumor as mature cystic teratoma containing adipose tissue (mesodermal origin), mucous gland acini (mesodermal origin), cystic areas lined by ciliated pseudostratified columnar epithelium (endodermal origin), and keratin debris (ectodermal origin). Thus, derivatives of all three germ layers were present in this case.

Teratomas are classified as mature, immature and malignant teratomas. Mature teratomas mainly contain mature elements such as cartilage, squamous epithelial cells, glands, mucosal tissue, and neural elements. Immature teratomas have a tendency to recur and are aggressive tumors, comprising primitive, undifferentiated components that resemble fetal tissues. Malignant teratomas are derived from the yolk sac or endodermal sinus, and especially, malignant teratomas, along with the high levels of serum  $\alpha$ -fetoprotein, are associated with a poor prognosis.<sup>[8,27]</sup>

There are two dominant theories regarding the origin of intraspinal teratomas-the dysembryogenic theory and the misplaced germ cell theory.<sup>[28,29]</sup> According to the dysembryogenic theory, spinal teratomas arise from the pluripotent cell and that in a locally disturbed developmental

environment, these pluripotent cells differentiate chaotically. When such disordered development occurs in a primitive streak or a caudal cell mass, a spinal teratoma forms.<sup>[27,30]</sup> Another is the misplaced germ cell theory according to which certain pluripotent primordial germ cells of the neural tube that get misplaced during migration from the yolk sac to the gonad, lead to spinal teratoma formation.<sup>[29]</sup> In adult intraspinal teratomas, which rarely present with significant dysraphism, the misplaced germ cell theory is likely to be more feasible.<sup>[8,23,28,31,32]</sup> The mature teratoma in our case may support the idea of a tumor actually arising from misplaced pluripotent primordial germ cells.

Long-term follow-up is required to determine whether residual remnants of the lesion can cause a recurrence. Due to the extremely low incidence of adult mature spinal teratoma and the limited knowledge of the disease, adjuvant therapy for such teratomas remains controversial.<sup>[24]</sup>

## Conclusions

Intradural extramedullary teratoma is a rare tumor. The diagnosis is based on the intra-operative and the histopathological examination. Total surgical excision is the primary treatment modality. Long-term follow-up is required to rule out recurrence of the tumor.

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