Mature cystic teratoma of spinal cord in an adult. A rare tumour

Rajneesh Misra, Sushil Kumar, Sandeep Sharma
Mature cystic teratoma of spinal cord in an adult. A rare tumour

Rajneesh Misra¹, Sushil Kumar², Sandeep Sharma³

¹ Robert Jones and Agnes Hunt Royal Orthopaedic Hospital, Oswestry, UK
² St. Stephens Hospital, Tis Hazari, Delhi, INDIA
³ Safdarjung Hospital, Ansari Nagar East, New Delhi, INDIA

ABSTRACT
Aim: Reporting a rare case of mature cystic teratoma of spinal cord in an adult
Background: Teratomas of the central nervous system are rare lesions accounting for less than 0.5-2% of all CNS tumors. Most of these are found in pediatric age group and favor sellar - suprasellar, pineal and sacro-coccygeal regions. Their occurrence in spinal cord in an adult is incredibly rare.
Case presentation: A 22-year-old male presented with low back ache and weakness of both lower limbs. Clinical examination and radiological workup revealed presence of a cystic lesion causing diffuse enlargement of the cord from L1 to L5-S1. Histopathological examination of the excised lesion revealed it to be teratoma.
Conclusion and clinical significance: Mature teratoma of the spinal cord in adults is extremely rare occurrence. However, this diagnosis should be kept in mind when evaluating a cystic lesion of cord even in adults. Maximal safe resection of the lesion results in prolonged deficit free survival.

BACKGROUND
Teratomas of the central nervous system are rare lesions. Barring the sacro-coccygeal region, their occurrence in spinal cord is a rarity. They constitute only 0.5% of intraspinal tumors.

CASE PRESENTATION:
A 22-year-old male had presented with complaints of weakness of both lower limb in the last three months. He had difficulty in clearing the feet off the ground. This was associated with altered sensation in the left leg in a below knee distribution where he was not able to feel the temperature of water properly when bathing. He had a history of low back ache on and off for 4 years which was relieved by rest and analgesics. There were no ‘red flags’ till his presentation with weakness. He denied any history of trauma, lumbar puncture, bowel or bladder incontinence or fever. He was not forthcoming about his sexual history probably due to cultural reasons.

On examination, his vitals were normal, and the local examination of spine did not reveal any stigmata of spinal dysraphism. There was no
deformity. The power in both ankle dorsiflexors and great toe extensors was 3/5. It was largely unaffected in all other muscle groups. Sensory examination revealed almost complete loss to touch, pain, and temperature in left L4 to S1 distribution and by about 25-30% in S2 to S4 distribution. Bilateral knee and ankle jerks were absent, and planters were mute. MRI imaging revealed a large irregular intradural lesion of spine extending from L2 to L5-S1 junction which was hypointense on T1 and hyper intense on T2 with irregular enhancement on contrast (Fig. 1).

**Figure 1.** Preoperative MRI shows the lesion extending from upper border of L2 vertebra to lower border of L5. It has typical cystic appearance, hypo intense on T1 (A & D) with enhancement of walls on contrast (B & E). The contents are hyperintense on T2 (C & F) along with a syrinx in the cord. The lesion itself is pointed with hollow arrows.

The patient underwent L1-L5 laminectomy. On dural opening, a thick-walled lesion was found adherent to the conus and cauda roots. On opening the cavity, greenish pus-like fluid was drained. The wall was dissected piecemeal, and a small residual was left over one of the roots. The intraoperative picture was indicative of an epidermoid cyst. Histopathological picture showed presence of all three germ cell layer derivatives indicative of a mature teratoma. Post operative period was complicated by presence of CSF leak which abated after removal of drain, pressure dressing and keeping the patient in prone position for 3 days. Before discharge on 10th day, the patient’s motor status had improved to 4/5 in both ankle dorsiflexors and great toe extensors. He had a residual sensory loss of about 50% in left L5 and S1 distribution. He has been followed up for last seven years and he has recovered a power of 5/5 in all muscle groups with minimal sensory loss in left S1 distribution. Follow up MRI showed presence of a residual lesion at the tip of conus which was mildly hyperintense on T1 with no contrast enhancement and hypointense on STIR (Fig2). This is consistent with fatty tissue which may have been the small fragment that was left attached to one of the roots intraoperatively.

**Figure 2.** MRI image at eight years follow up shows residual lesion (marked with hollow arrows) opposite the lower half of L3 vertebra which is hyperintense on T1(A&D) and hypointense on STIR (B & E) indicating fatty tissue. There is no evidence of syrinx.

**DISCUSSION**

The spinal tumors are classified according to their location. The IDEM lesions constitute about 55% of all spinal tumors. The most common IDEM tumors in
adults are nerve sheath tumors and meningioma [1].

Virchow reported the first spinal teratoma. This occurs more frequently in children with spinal dysraphism and is associated with various malformations like spina bifida, tethered cord, and split cord malformations of various types [2-7]. Sloof et al in their series of 1322 patients of spinal cord tumors, identified only 2 patients with teratoma [8]. Al-Sarraj et al had seven cases with a histological diagnosis of teratoma in a series of 25,000 cases over a period of 15 years [9]. Thus, spinal cord teratomas are in general very rare. Moreover, they occur far less frequently in adults.

Spinal teratoma in adults can have variable presentation and in general have a set of presenting symptoms like other IDEM lesions. However, there is a significant alteration in frequency of these symptoms compared to IDEMs in general. Usually, there is nothing in presentation that points specifically to the diagnosis. In our case, the patient presented with syndrome suggestive of a conus-cauda lesion. Poeze et reviewed 31 cases of intramedullary teratomas and found motor dysfunction to be the most common presentation (71%), followed by changes in reflexes, sensory changes, urinary changes, and pain [6]. Far less common were sexual disturbances and anal sphincter disturbances. On the other hand, a prospective study in 107 patients of IDEM tumors by Tarantino et al found pain to be the most common presentation followed by sensory deficits. The motor deficits at presentation were distant third (12%) [10].

Specific diagnosis of teratomas on imaging is difficult. They, in general have a picture like other IDEM tumors. On MRI, they appear lobulated or cystic, with variable signal intensity and enhancement consistent with the different solid and cystic components of the tumor [11,12]. CT imaging is nonspecific and reveals differences in densities within the tumor consistent with different tissue types [13]. The role of imaging is more for preoperative planning and the decision regarding location of the tumor in relation to the cord rather than a definitive diagnosis.

Ultimately, the diagnosis of teratoma rests on histopathological demonstration of all the three germ layers in the specimen. Currently there are two proposed theories for the origin of spinal teratoma, the misplaced germ cell theory and the dysembryogenic theory [14, 15]. Regardless of the origin, the prognosis of teratoma is dictated by the classification into mature, immature, and malignant. The mature ones have an overall benign course once resected. However, the malignant teratoma as the name suggests, have an aggressive and stormy course with poor prognosis. The primary mode of treatment is surgical resection, and the goal is decompression of the neural tissue without causing further deficits. However, complete resection is a realizable goal in about 50% of cases and should always be attempted with patient safety in mind [16]. However, many authors suggest not attempting a complete resection citing the logic that these are slow growing tumors affording a large window period for picking up a recurrence and that the rate of recurrence with partial and complete resection is very similar (11% vs 9%) [17,18].

The overall prognosis for mature teratoma of spine is good following surgical resection with most patients reporting either improvement or stabilization of symptoms. However, most case reports [3,6] have a short follow up. The patient in our case was followed up for approximately 8 years and showed improvement of all symptoms except mild sensory abnormality in left S1 dermatome. The rarity of these lesions in adults makes it difficult to design a long-term study of disease progression. Currently, there is no evidence supporting any role for chemotherapy or radiotherapy for mature teratoma following resection [19].

CONCLUSION AND CLINICAL SIGNIFICANCE

Spinal mature teratomas in adults are very rare. The clinical presentation is like any other IDEM tumor or conus-cauda lesion. Imaging modalities also do not offer any specific distinguishing features from other similar lesions in the region. However, this differential diagnosis must always be considered when evaluating a radiological picture of cystic lesion of the spinal cord. The treatment rests on maximal safe surgical resection with an attempt for complete resection. These patients are likely to have a prolonged near deficit free course even in the long term.

List of Abbreviations

MRI: Magnetic resonance imaging
T1WI: T2 Weighted image
T2WI:T1 Weighted image
IDEM: Intradural extramedullary
CT: Computer tomography
CSF: Cerebrospinal Fluid
STIR: Short tau inversion recovery

REFERENCES