

Spinal Intradural Extramedullary Mature Cystic Teratoma at Cervical Location in an Elderly Person: A Rare Case Report with Review of Literature

Case Report

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Abstract

Spinal intradural extramedullary teratoma is a rare condition in adults and, at cervical spinal location, it is extremely rare. We reviewed many literatures and PubMed advanced search showed 11 results of intradural extramedullary teratoma in adults (above 18 years) which included nine independent case reports and three cases in a case series. We also found only one intradural extramedullary teratoma at cervical spinal position. Here, we reported a rare case of an adult onset intradural extramedullary teratoma in the cervical spinal cord with no evidence of spinal dysraphism and without any history of prior spinal surgery. The patient was a 70-year-old female whose chief complaints were neck stiffness and numbness at her left upper extremity. Magnetic resonance imaging of spine revealed a cystic intradural extramedullary mass opposite the 2nd cervical vertebra. The patient was operated and the mass was totally excised. The resected tumour was diagnosed histopathologically as a mature cystic teratoma. The patient's symptoms were improved following the surgery.

Introduction

Teratomas are types of tumours containing tissues from all three germ layers and result from ectopic growth of these tissues [1,2]. Based on the degree of differentiation, teratomas may be classified as mature, immature and malignant teratoma [3]. The incidence of spinal teratoma is rare, only 0.15-0.18% of spinal tumours are intra spinal teratomas [4]. In paediatric patients, 5-10% of spinal tumours are intra spinal teratomas [5-7], whereas the incidence among adult patients is significantly lower than that is observed in childhood and infancy [1,8-13].

Case Report

A 70 years old lady, who was apparently healthy before, presented with neck pain, stiffness along with progressive numbness and weak-

ness in her left arm and hand for 2 months. There was no history of any congenital deformity, spinal injury, surgery or any other spinal procedures like lumbar puncture. The physical examination revealed that, there were no sensory or motor deficits in the extremities, no palpable mid line spinal displacement. Upon neurological examination, she demonstrated normal physical reflexes. No cutaneous abnormalities or dermal sinus tract were found. The lab parameters were unremarkable. The Magnetic Resonance Imaging (MRI) of spine revealed an intradural extramedullary lesion measuring about 9.7mm x 7.2mm, within the left side of the spinal canal at 2nd cervical vertebral level compressing the cord to the right. The lesion showed T1WI and T2WI isointense signal changes. On post contrast, the lesion showed strong homogeneous enhancement (Figure 1,2). Radiological diagnosis was most likely schwannoma and differential diagnosis revealed meningioma. Surgical intervention was planned and patient underwent total



resection of tumour by means of a C2-C3 laminectomy. Through an incision to dura via mid line incision, a yellow oval fatty cyst was observed. Separation of the capsule and total removal of the tumour was done. The histopathological examination of the excised mass revealed the presence of elements from 2 germ cell layers (ectoderm and mesoderm). Any tissue of endodermal origin was not found. Under light microscope, sections showed fibrous tissue and bundles of spindle shaped Schwann cells (Figure 3). These revealed cystic areas containing fragments of keratin materials (Figure 4,5). Few hair shafts with foreign body type multi-nucleated giant cells were also seen (Figure 6,7). Areas around fatty tissue, fibrous tissues, mature muscle tissues were also noted (Figure 8-10). No caseating granuloma or malignancy were seen. The final histopathological diagnosis was spinal mature cystic teratoma with foreign body granuloma. Following surgery, her symptoms were gradually improved and no further neurological deterioration was observed during 6 months of follow-up period.

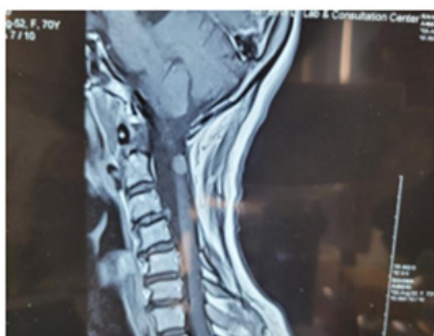


Figure 1: Sagittal T1- weighted MRI contrast enhanced scan revealing an intradural extramedullary lesion at C2 vertebral level.

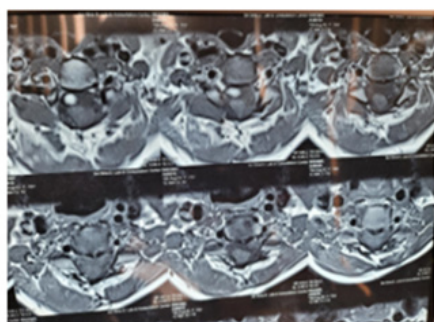


Figure 2: Axial T1 weighted MRI image showing an isointense lesion in left side of the spinal canal at C2 vertebral level compressing the cord to the right.

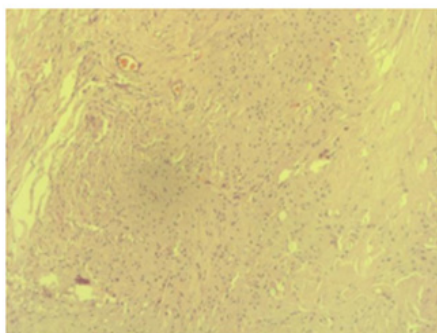


Figure 3: Section shows Schwann cell (representing ectodermal tissue).

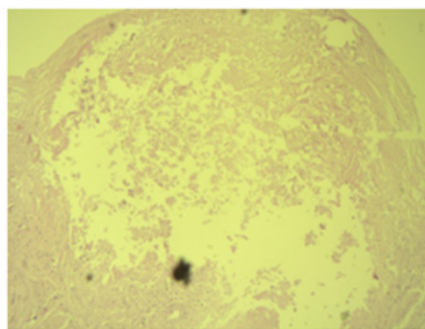


Figure 4: Section shows keratin material (representing ectodermal tissue).



Figure 5: Section shows keratin material (representing ectodermal tissue).

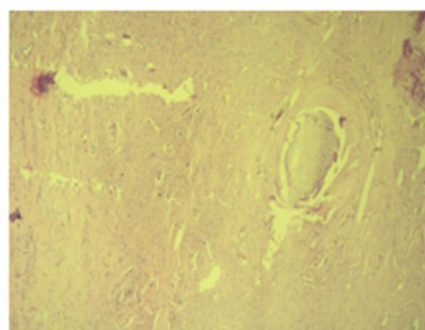


Figure 6: Section shows hair shaft (representing ectodermal tissue).

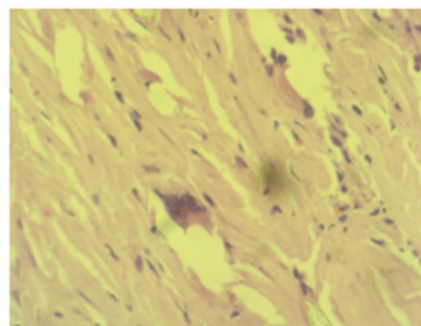


Figure 7: Section shows multi-nucleated giant cells (representing mesodermal tissue).

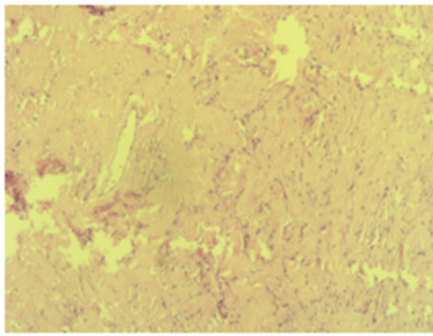


Figure 8: Section shows blood vessel with fibrous tissue (representing mesodermal tissue).

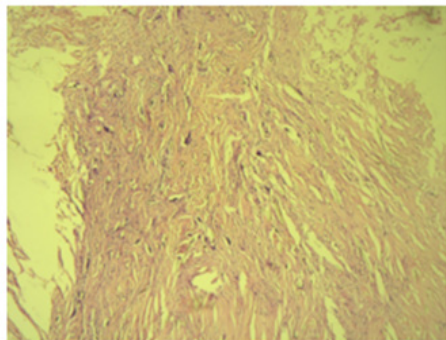


Figure 9: Section shows fibrous tissue (representing mesodermal tissue).

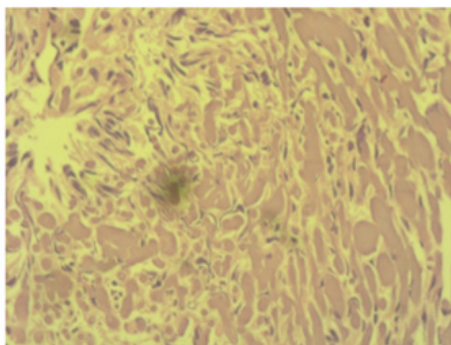


Figure 10: Section shows skeletal tissue (representing mesodermal tissue).

Discussion

The spinal teratoma is an uncommon disease, which accounts for only 0.1-0.5% of all spinal tumours [1,14]. The first case of this condition was described by Virchow in 1863 [15] and Gowers and Horsley described another case 25 years later [16]. Only few series of studies on this condition have been reported in medical literature. Spinal intradural extramedullary teratoma is a rare entity in adults, more common in children and often associated with spinal dysraphism [17]. The association of teratoma and spinal malformation such as spina bifida, partial sacral agenesis, hemi vertebrae, myelomeningocele, tethered cord syndrome diastematomyelia has been described in the literature [1,18-21]. Besides that, several authors reported that a history of trauma or spinal surgical procedures often precedes the clinical presentation of this condition in adult case [22,23]. We reviewed the literatures and PubMed advanced search showed 11 results of intradural extramedullary teratoma in adults (above 18 years) which includes nine independent case reports and three cases in a case series (Table 1). We reported a rare case of intradural extramedullary teratoma at cervical spine in an elderly woman (70 years) without any spinal dysraphism or h/o spinal procedures. Although, sacro-coccygeal teratoma in new

born babies have frequently been reported, in adult cases of spinal teratoma, the thoraco-lumbar region is most commonly affected, particularly in the area of conus medullaris [1,18-20]. However, in our case, the lesion was at cervical spine which is very rare, only 5 cases of cervical intradural teratoma in adults have been reported in literature till present and all the cases were female and among them 4 cases were intra medullary and only 1 case was extra-medullary (Table 2).

The clinical features, including weakness of the extremities, sensory change and reflex abnormalities, are related to the location of the tumour [18]. In the literatures, most of the cases have presented with these symptoms with regard to the clinical manifestations of the tumour in our case patient had neck stiffness and numbness of left upper extremity [24-29]. In the diagnosis of spinal teratomas, MRI is regarded as gold standard diagnostic tool that can detect the location of teratoma and, consequently, the degree of spinal cord involvement [30-32]. The MRI finding of intradural extramedullary tumours include displacement of the cord to the contralateral side of the thecal sac, widening of the sac available for CSF above and below the tumour and a sharp demarcation between the tumour and cerebrospinal fluid [33]. In our case, MRI scan revealed a well-defined intradural extramedullary lesion within the left side of spinal canal, located opposite body of C2 vertebral level, compressing the cord toward right, accounting for neck stiffness with numbness and weakness of left upper extremity. The pre operative radiological diagnosis of spinal teratoma is difficult as the MRI features cannot determine with certainty the differential diagnosis between teratoma and other extramedullary lesions like nerve sheath tumours (neurofibromas and schwannomas) and meningiomas [15]. Surgery followed by histopathological examination is must to confirm the diagnosis of intra spinal teratoma.

The diagnosis of teratoma depends on the histopathological identification of the tissue representing three germinal layers (ectoderm, mesoderm and endoderm) [34]. The current classification states that a teratoma is a tumour that is typically but not always composed of derivatives from all 3 primitive germ layers [18]. Li et al. [35] analysed the literature and found in a number of cases, only two of the three germinal layers were observed and attributed this to the fact that the derivatives of one or two of the layers had grown over the others [1,35,36]. 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, comprising primitive, undifferentiated components that resemble fetal tissues, have a tendency to recur and are aggressive tumours. Malignant teratomas are derived from the yolk sac or endodermal sinus, especially, malignant teratomas, along with high levels of serum alpha fetoprotein, are associated with poor prognosis [19,37].

There are two dominant theories regarding the origin of intra spinal teratomas. The first is the dysembryogenic theory, and the second is the misplaced germ cell theory [38,39]. The dysembryogenic theory indicates that spinal teratomas arise from pluripotent cells, 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 [37,40]. The misplaced germ cell theory suggests that certain pluripotent primordial germ cells of the neural tube are misplaced during migration from the yolk sac to the gonad, thus resulting in spinal teratoma formation [39]. In adult intra-spinal teratomas, misplaced germ cell theory is likely to be more feasible [38]. The mature teratoma in our case may support the theory of tumour actually arising from misplaced pluripotent primordial germ cells. The primary treatment modality for symptomatic patients is total surgical resection. However, these tumours are difficult to resect completely due to adhesion to the surrounding tissues found in about 50% cases [1,19,20,41]. Removal of as much of the pathological tissue is recommended if complete removal of tissues is not possible to preserve surrounding neural tissues since subtotal resection increases chance of recurrence [20].



Table 1: Review of intradural extramedullary teratoma cases in adults (18-85 years).

Author/Year	Number of Cases (n)	Mean Age (in years)	Sex	Site	Position	Associated Abnormality
Kumar et al. [24]	01	19	F	IDEM	T5, T6	Spinal dys-rhaphism
Stevens et al. [23]	01	85	M	IDEM	L1, L2	Absent
Sung et al. [18]	01	38	M	IDEM	T11, T12, L1	Absent
Liu et al. [25]	003	Individual cases not found	N/A	N/A	N/A	N/A
Bouaziz et al. [26]	01	38	F	IDEM	Cervical Spinal cord (on both side of foramen magnum)	Absent
Vanguardia et al. [27]	01	41	M	IDEM	T12/L1	Absent
Agay AK et al. [28]	01	20	M	IDEM	D11, D12	Absent
Kafadar C et al. [29]	01	48	F	IDEM	L2-L4	Tether cord (conus medullaris at L4-L5 levels), diastematomyelia, block vertebra and spina bifida occulta
Shrestha A et al. [30]	01	19	M	T12-L2	IDEM	Absent

*IDEM=Intradural Extramedullary.

Table 2: Review of intradural teratoma cases (both intramedullary and extramedullary) at cervical location in adults.

Author/ Year	Number of Cases (n)	Mean Age (in years)	Sex	Site	Position	Associated Abnormality
Kubie LS et al. [31]	1	27	F	C3-C4	IDIM	Absent
Ak et al. [19]	1	43	F	C2-C3	IDIM	C3 spina bifida, c5 level nodule
Makary et al. [20]	1	46	F	C1-C2	IDIM	C1-C2 dysraphic congenital spinal malformation
Ghostine et al. [32]	1	65	F	C1-C2	IDIM	Absent
Bouaziz et al. [26]	1	38	F	At cervical spinal cord and bulbo-medullar junction.	IDEM	Absent
Present	1	70	F	C2	IDEM	Absent

*IDEM=Intradural Extramedullary; IDIM=Intradural Intramedullary.



Recurrence of the tumor leading to the reappearance of symptoms takes a longer time due to the tendency of these tumors to grow slowly [20]. Total and subtotal resections seem to have similar recurrence rates (9% and 11%), respectively; thus, some authors do not recommend aggressive resection which could lead to neurological deficit [1,19,42]. To prevent the cystic contents from spilling into the intradural space which may lead to aseptic chemical meningitis with or without obstructive hydrocephalus, care should be taken during surgery [22,37,43]. For densely adherent lesions, intraoperative electrophysiologic monitoring, which signals when the dissection should be stopped, can be extremely useful in resection of the lesions, to prevent permanent neurological damage, especially with lesions in higher spinal levels [44-46]. Long-term follow-up of the cases is required to rule out possible recurrence with repeated MRI imaging [34,35,47]. There is no definitive evidence of additional benefits of radiotherapy and chemotherapy in these tumors [35,41,48].

Conclusion

Intradural extra medullary teratomas are rare tumours in adult and, at cervical location they are extremely rare. The diagnosis is based upon histopathological findings. Total surgical excision is preferred; however, the total and subtotal resections seem to have similar recurrence rates. A long term follow up should be done to rule out recurrence of the tumour.

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