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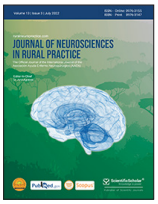

Case Report

Ossified upper thoracic intradural mature teratoma in a teenager – A technical case report with special emphasis on surgical strategy

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ABSTRACT

Spinal teratomas are heterogeneous neoplasms and are extremely rare in the upper thoracic spine. They are sub-classified as mature, immature, or malignant. They may be calcified or rarely ossified; the latter posing a major surgical challenge due to surgical difficulties in safe removal. Clinico-radiologico-pathological and operative experience of ossified spinal intradural mature teratomas is extremely rare. We present a case of ossified upper thoracic intradural mature teratoma managed by microsurgical drilling and resection under neuromonitoring.

Keywords: Ossified, Thoracic, Intradural, Mature teratoma, Spinal cord neoplasm, Children

INTRODUCTION

Spinal teratomas are rare, slow growing, disorganized, and heterogeneous collection of tissues derived from all three primitive germ layers. Spinal intradural mature teratomas are extremely rare in the upper thoracic location.[1] Intradural ossification though often seen in benign tumors (meningioma, Schwannoma, etc.), infection (arachnoiditis), or hemorrhage has rarely been reported in mature teratomas.[2,3]

CASE REPORT

An 18-year-old boy presented with slowly progressive weakness and numbness of both lower limbs for the past 3 years, and chronic constipation with difficulty in passing urine for the past 5 months. The patient was catheterized periurethrally for continuous dribbling of urine 5 months back and is bedridden since then. There was no history of spine surgery or previous lumbar puncture. Clinical examination showed severely spastic paraparesis and complete sensory loss (all modalities) below T2 dermatome and lax anal tone. There were no neurocutaneous markers or cutaneous stigmata of occult spinal dysraphism.

Pre-operative CEMRI spine [Figure 1a.1-5] showed a rim enhancing ventrally placed cystic intradural lesion at D2-3 vertebral body level without any diffusion restriction. Another heterogeneous lesion was noted posteriorly at D3-4 vertebral body level causing thinning of thoracic cord due to impingement between the larger ventral and smaller dorsal lesion. Pre-operative computed tomography (CT) [Figure 1b.1-4] showed a non-uniformly thick continuous crescent-shaped ossification (>1000 Hounsfield unit [HU]) in the dorsal portion of spinal canal.

Provisional diagnosis of a heterogeneous, ossified intradural benign spinal tumor such as meningioma or nerve sheath tumor; or a ruptured neurenteric cyst/dermoid with ossification of spilled out material; or presence of dual pathologies was kept.[4] After D1-D4 laminectomy in prone position, hypervascular but normal looking duramater was opened in midline. There were dense but dissectable adhesions between the inner surface of the duramater and the underlying ossified tissue throughout the extent of laminectomy. Intradural exposure was extended beyond the cranial and caudal limits of ossification [Figure 2a] which was yellowish-white, bony hard, and not breakable even by ultrasonic bone scalpel (SonaStar, Misonix, USA). Few hairs embedded in the underlying ossified lesion are shown in [Figure 2b]. Using pneumatic drill (Midas Rex, AM8 cutting drill tip, Medtronic, USA) under copious saline irrigation, the ossified portion of the lesion was thinned out and then removed using Kerrison punch from below upward [Figure 2c-e]. The ossified portion of the lesion though was impinging and encasing the dorsal and lateral surfaces (right > left) of the thoracic cord but was separate from the cord

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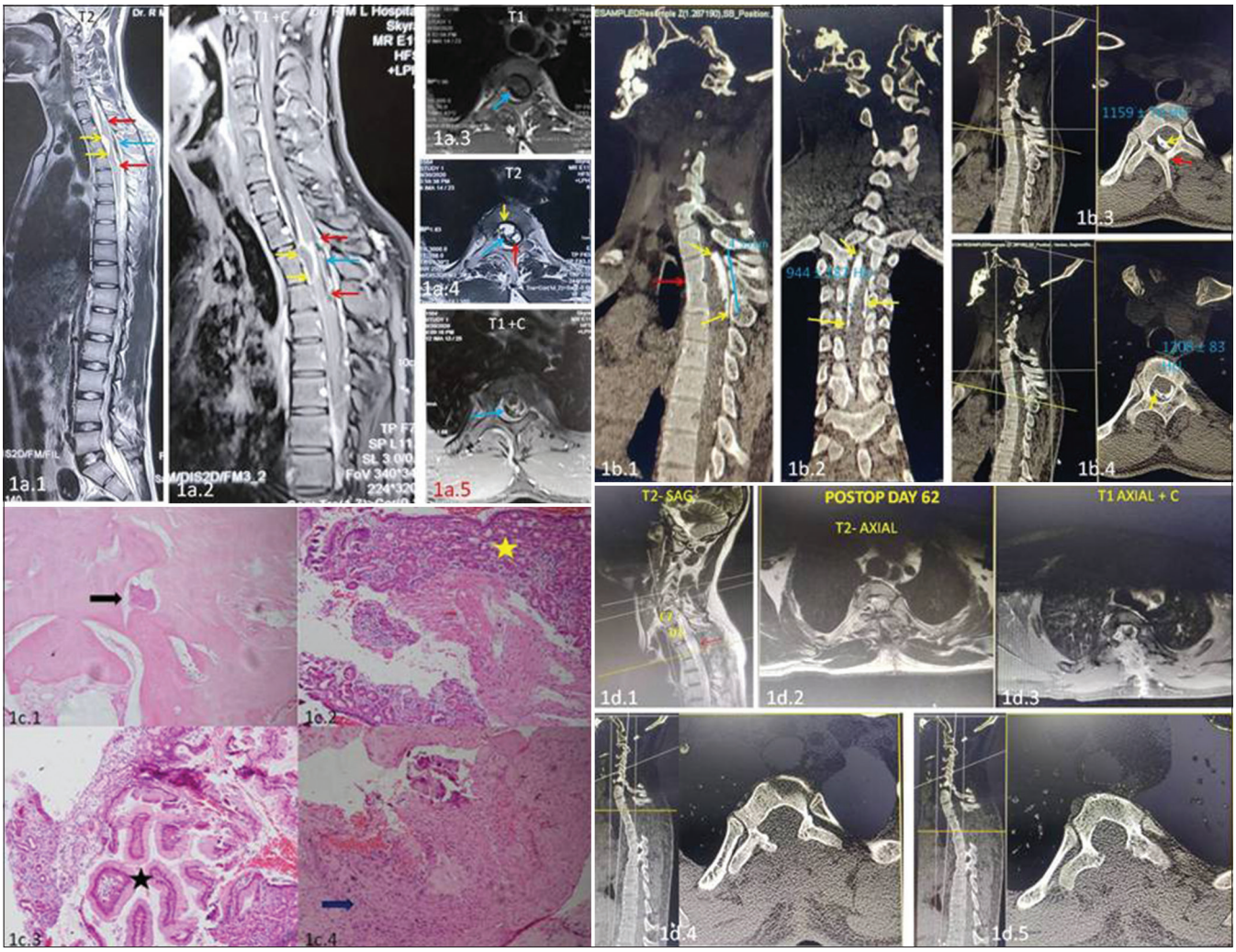

Figure 1: (1a.1-5) CEMRI dorsal spine with whole spine screening showed a T1 hypointense, T2 hyperintense, rim enhancing ventrally placed intradural cystic lesion at D2-3 vertebral body level of size 2.2 × 0.8 × 1.0 cm without any diffusion restriction (yellow arrows). Another heterogeneous lesion of size 1.9 × 0.3 × 0.7 cm was noted posteriorly at D3-4 vertebral body level (red arrows) causing thinning of thoracic cord due to impingement between the larger ventral and smaller dorsal lesion. An irregular vertical hypointensity (both on T1 and T2) was also noted in the dorsal part of spinal canal (sky blue arrow). D2-4 congenital block vertebra with rudimentary intervening discs, cervicodorsal scoliosis, and diffuse spinal pan meningeal enhancement were also seen. (1b.1-4) CT Cervicodorsal spine showed a thick continuous crescent-shaped (concave anteriorly) ossification (sky blue dots indicating HU value) in the dorsal portion of spinal canal (yellow arrows) without any attachment to vertebral bone, along with segmentation anomalies of anterior and posterior elements of D2-4 vertebrae (red arrow) and cervicodorsal scoliosis. (1b.3-4) Yellow colored line level marker is seen in sagittal image on the left with its corresponding axial image on the right. (1c.1-4) Histopathological examination showed (1) bony trabeculae lined by osteoblasts with interspersed squamous epithelium (black arrow). (2-3) In addition, fragments of gastric mucosa (yellow asterisk) and intestinal mucosa (black asterisk) were identified. (4) There is evidence of focal arachnoid cell proliferation (blue arrow). No immature elements or evidence of malignancy was seen. (1d.1-5) Follow-up imaging on post-operative day 62 with yellow colored line level marker in sagittal images. (1,2,3)- CEMRI Cervicodorsal spine showing good decompression of thoracic cord with focal myelomalacia of thoracic cord (red arrow) at the level of D2 vertebral body. Good decompression of ventral cyst was evident. (4,5)- CT cervicodorsal spine showing upper thoracic vertebrae laminectomy defect and dorsal spinal canal totally free off any bony lesion.

by its pia mater which was yellowish-white, thickened, and intact at most of the places except at the place, where bony crescent was deeply embedded into the cord on the right side [Figure 2f]. After excising the bony portion, a cystic lesion was seen ventral to cord on the right side at D2-3 body level – it was initially punctured and transparent gel-like fluid was aspirated from the right side; its wall was then dissected at the maximum possible and excised [Figure 2f-g]. Cord was lax at the time of closure [Figure 2h]. Motor evoked potentials were normal in the upper limbs, but not recordable

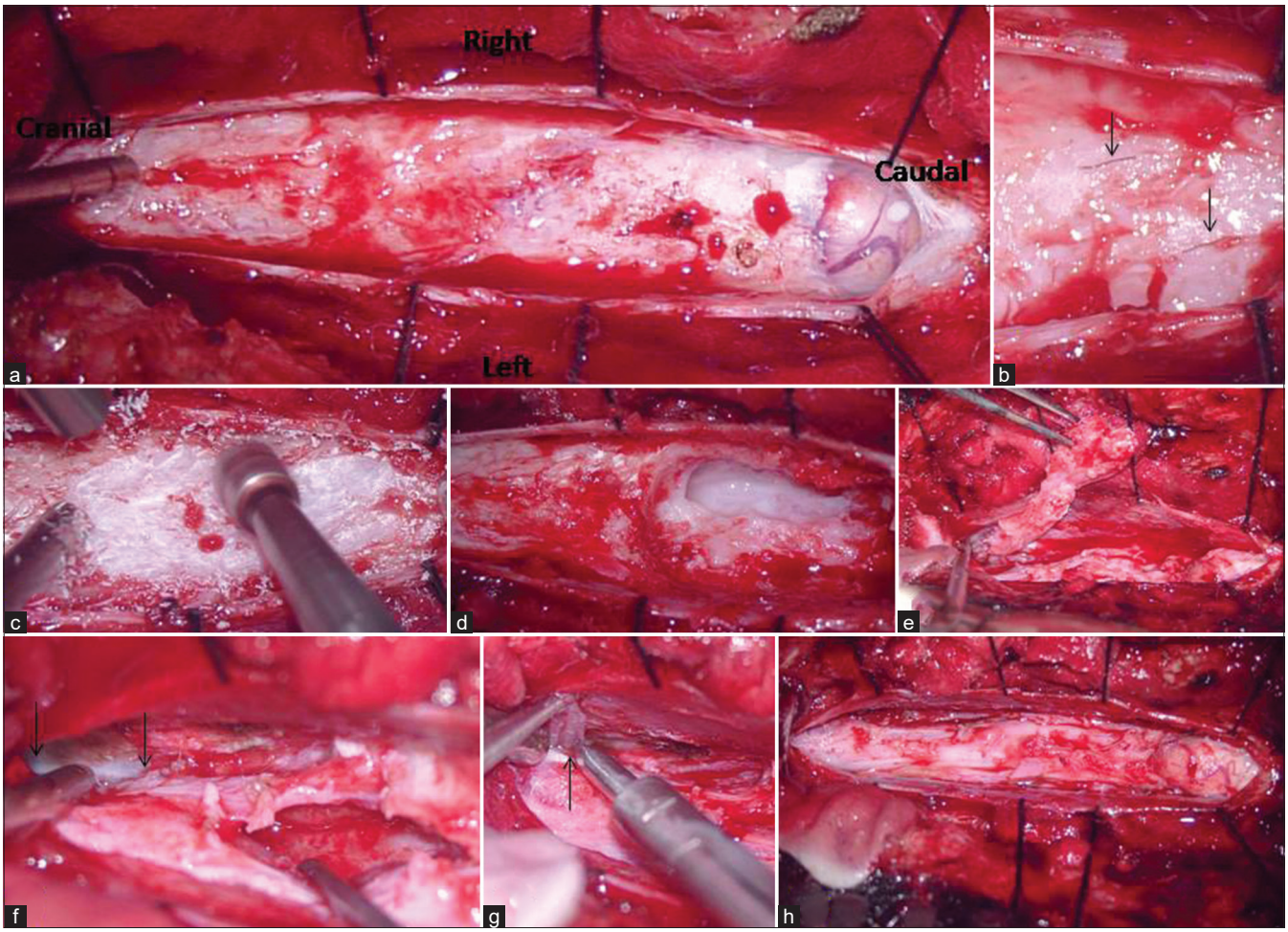

Figure 2: (a-h) Intraoperative images showing (a) intradural exposure (after adhesiolysis) beyond the cranial and caudal limits of ossified tissue which was yellowish-white, bony hard, not breakable even by ultrasonic bone scalpel; (b) few hairs embedded in the ossified lesion were seen; (c-e) the ossified portion of the lesion was thinned out using cutting drill and then removed using Kerrison punch from below upward; (f) the ossified portion of the lesion though was impinging and encasing the dorsal and lateral surfaces (right > left) of the thoracic cord but was separate from the cord by its pia mater which was yellowish-white, thickened, and intact at most of the places except at the place where bony crescent was deep on the right side; a cystic lesion was seen ventral to cord at D2-3 body level on the right side (marked by black arrows) - it was initially punctured and clear watery gel-like fluid was aspirated; (g) wall of the cystic lesion was then dissected at the maximum possible and excised; and (h) cord was lax at the time of closure.

in lower limbs throughout the procedure. Furthermore, D-wave was not recordable. Post-operative recovery was uneventful. Post-operative serum and cerebrospinal fluid germ cell tumor markers (alpha-fetoprotein, beta-human chorionic gonadotropin, and lactic acid dehydrogenase) were normal. Histopathological examination [Figure 1c.1-4] revealed features consistent with mature teratoma with ossification. His lower limbs tone was still high (reduced compared to pre-operative status) without any improvement in motor power along with complete sensory loss below nipples and complete loss of bowel and bladder sensations at 8 months follow-up without any radiological recurrence [Figure 1d.1-5].

DISCUSSION

Spinal teratomas though more commonly present in childhood (first or second decade of life) can rarely present in adults and elderly. They can be extradural or intradural (extramedullary, exophytic intramedullary, or intramedullary).[5]

Although the exact pathogenetic mechanism of spinal teratomas remains unclear; misplaced germ cell theory (misplacement of pluripotent primordial germ cells during migration from yolk sac to gonadal ridges) and dysembryogenic theory (teratoma arising from chaotically differentiated pluripotent cells in primitive streak or

caudal cell mass due to locally disturbed developmental environment) are the two well accepted theories to explain their etiopathogenesis.[1,5-8] Association of spinal teratomas with congenital spinal anomalies such as spina bifida, partial sacral agenesis, hemivertebrae, myelomeningocele, tethered cord syndrome, and diastematomyelia is well described in Literature.[9]

Their clinical presentation is not specific and resembles that of other spinal space-occupying lesions of similar anatomical locations.[5,10,11] Magnetic resonance imaging (MRI) being the neuroimaging tool of choice shows a heterogeneous solid-cystic tumor containing fat and calcium.[5] Peculiar features noted on neuroimaging include simultaneous presence of intratumoral calcification (hypointense on T1 and T2) and adipose tissue (hyperintense on T1 and hypointense on T2 and fat suppression sequences) and no contrast enhancement (except in patients with history of recurrent aseptic meningitis). CT scan is indicated in patients having intra- or juxta-lesional calcification/ossification and congenital bony anomalies. However, all these neuroimaging findings are only suggestive but not diagnostic of teratoma, but are extremely helpful in sequencing of differential diagnoses list.[5]

With the early surgery as the treatment of choice, maximal possible safe resection while preserving the surrounding neural tissue using intraoperative electrophysiological monitoring is the goal.[9,11] Complete excision with intact capsule though feasible in extradural and extramedullary locations is unlikely in intramedullary teratomas.[5] Intraoperative spillage of tumor contents should be avoided, and surgical field should be thoroughly irrigated with normal saline, especially before dural closure to prevent acute post-operative complications such as aseptic meningitis or hydrocephalus.[7] Maximum possible excised tumor tissue should be collected to be sent for histopathological examination and the complete specimen should be analyzed to detect malignant cell populations interspersed between islands of benign tissue.[1] Thus, serial sectioning is essential.[11] Extramedullary mature teratomas have higher rates of complete resection compared to their intramedullary counterparts due to cord invasion and absence of cleavage plane throughout in the latter.[5] Long-term symptomatic recurrence rates following total (9%) and subtotal (11%) resection of mature teratomas have been found to be nearly similar.[7]

Making a diagnosis of spinal teratoma is based purely on the histopathological features (gold standard) as it is difficult to differentiate it from other lesions based on clinical, pre-operative neuroimaging, and/or intraoperative findings.[5,9] Although demonstration of derivatives from all the three germ layers is needed for making the diagnosis of a teratoma; it cannot be ruled out if derivatives of only two germinal layers are demonstrated.[9]

Ossification and calcification can be differentiated radiologically (measure radiodensity - Hounsfield Units) and histopathologically (more precisely) as calcification is mere deposition of calcium salts in soft tissues recognized by presence of psammoma bodies, whereas ossification is a process of new bone formation.

Prasad and Divya,[5] in their study of 146 cases of adult spinal teratomas, have reported overall recurrence rate of 6.1% (n = 9) over a mean follow-up of 37 months (2 weeks to 208 months). All the recurrences were managed by re-surgery and no malignant changes were found in these recurred cases. Thus, recurrences though uncommon in mature teratomas are seen late due to extremely slow growth and are amenable to re-surgery.[5]

There is no role of adjuvant radiochemotherapy even in recurrent mature teratomas, this being strictly reserved for teratomas with immature or malignant components even after total resection.[5] Tumor markers do not have significant role in monitoring for recurrence of spinal teratomas especially the mature ones.[5] Long clinicoradiological follow-up for several years on annual basis is recommended in mature teratomas.[8]

We did literature search for massively ossified mature spinal teratoma cases and found only two such cases described in world literature [Table 1].[2,3] In pre-CT/MRI era, Azariah [2] has reported dense ossification in the thoracic spinal canal on pre-operative x-rays and ascending myelogram and confirmed it intraoperatively (bone removal using broad Northfield bone rongeur) and histopathologically (true bone with active hematopoietic marrow in tumor substance) in a 16-year-old girl with thoracic D4-9 intradural mature teratoma. Ijiri *et al*.[3] excised the L1-2 ossified mass totally using ultrasonic surgical aspirator, which on histological assessment showed presence of mature bone, cartilage, and mature adipose tissue with blood vessels (mesoderm derivatives). Although all the three germ layer derivatives could not be demonstrated, Ijiri *et al*.[3] still labeled it as mature teratoma with a clarification that the derivatives of mesodermal layer had overgrown the other two germ layers. The present case is the third case report in the English Literature reporting so massive intradural ossification in a mature teratoma along with its detailed surgical technique. Bone present in the lesion was surfacing dorsally on midline durotomy and there was no cleavage plane between the lesion and the thoracic cord tissue, thus suggesting it to be of exophytic intramedullary type. Intratumoral bone was so dense that both the ultrasonic surgical aspirator and bone scalpel failed to cut it. Bone removal was finally done using drill and Kerrison Rongeur alternating with microsurgical dissection (to free the bone chunks from surrounding cord tissue before their removal). Histopathological analysis demonstrated the derivatives of all the three germ layers.

Table 1: List of ossified mature spinal teratomas reported in the literature.

S No.	Author's name (Year)	Number of patients	Patient age and sex	Symptoms duration	Teratoma location and consistency	Neuroimaging	Associated anomalies	Extent of resection	Follow-up duration	Outcome
1.	Azariah *et al.* (1967)	1	16 Years/F	3 months	T 4-9, Intradural, solid mass	Spine X-rays- Almost complete absorption of the pedicles from T4 to T9 with a dense oval area of ossification within the spinal canal. Ascending myelography- obstruction at the lower border of T8.	Nil	Total	9 months	Improved
2.	Ijiri *et al.* (2009)	1	68 Years/F	4 months	L1-2, Intradural, solid mass	CT, MRI- Solid ossified mass at caudal tip of conus medullaris with hypointensity on T1 and T2 with peripheral enhancement	Nil	Total	36 months	Transient neurological deterioration-recovered later
3.	Present case (2021)	1	18 Years/M	36 months	T 2-4, Exophytic intramedullary, solid-cystic mass	CT, MRI- Heterogeneous irregular solid-cystic ossified mass impinging the cord with vertical hypointensity on T1 and T2 with diffuse spinal pan meningeal enhancement	D2-4 congenital block vertebra with rudimentary intervening discs, cervicodorsal scoliosis	Near total	4 months	No improvement

Non-improvement in the present case is likely to be due to poor pre-operative neurological status secondary to very late presentation (underwent surgery 3 years after symptoms onset).

Pre-operative neurological status and intraoperative handling of neural tissue while dissecting the ossified tumor portion microsurgically may be the key prognostic factors influencing the final neurological outcome.

CONCLUSION

Besides benign tumors, hamartoma, infection, or hemorrhage; teratoma should be considered higher up in the differential diagnosis of intradural ossified lesions especially in children.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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