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Case Report

Cervicomedullary junction mature teratoma with pulmonary differentiation and diastematomyelia in an adult – A rare case

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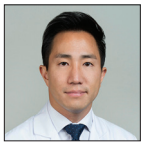

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ABSTRACT

Background: Intradural extramedullary teratomas in the cervical or cervicomedullary region are rare in adults.

Case Description: We report a symptomatic, mature teratoma at the cervicomedullary junction in a 52-year-old Hispanic female who also has a type I diastematomyelia in the thoracolumbar spine. The patient underwent surgical resection of the lesion with the resolution of presenting symptoms. Histopathology of the lesion revealed a mature cystic teratoma with pulmonary differentiation.

Conclusion: We discuss the case along with a review of pertinent literature and considerations with regard to the diagnosis, etiology, prognosis, and management of this unusual pathology.

Keywords: Cervicomedullary, Diastematomyelia, Mature, Teratoma

INTRODUCTION

Spinal teratomas are rare in adults (i.e., incidence is between 0.2% and 0.5% of all spinal tumors) and typically occur in the thoracolumbar region.[11,15] More commonly, these tumors are diagnosed in neonates and young children, typically involving the sacrococcygeal region where they are often associated with spinal dysraphism.[11,15] When found in adults, teratomas have been correlated with a history of trauma, prior surgery, or rare spinal dysraphism.[8] Here, we report a symptomatic intradural, extramedullary mature teratoma at the cervicomedullary junction (CMJ) in a 52-year-old Hispanic female with a remote history of having undergone posterior cervical surgery and also incidentally had a thoracolumbar diastematomyelia.

CASE DESCRIPTION

Clinical presentation

A 52-year-old Hispanic female with a history of type II diabetes mellitus presented with 2 months of occipital headaches, left upper extremity pain, and progressive left hemiparesis. There was no history of recent illnesses, trauma, or an underlying malignancy, but she had undergone posterior cervical surgery for a "mass" in Mexico at age 14. On examination, she demonstrated 4–/5 strength in both the left upper and left lower extremities along with a tender palpable soft-tissue mobile midline mass in the lumbar spine (i.e., consistent with a lipoma), and a well-healed 6 cm posterior transverse incision at the C2 level.

Computed tomography/Magnetic resonance (CT/MR) studies and differential diagnoses

The brain/cervical CT revealed a 2.0 cm lesion at the CMJ. The brain/cervical MR showed an intradural, extramedullary, and posterior mass (2.3 × 2.5 × 2.8 cm) with a cystic component located at the CMJ [Figures 1 and 2]. The MR T1 study without contrast showed that the entire lesion was hypointense with a subtle fluid-fluid level noted on axial images, while the T2 study revealed a hypointense lesion with a cystic component of similar intensity to cerebrospinal fluid [Figures 1 and 2]. Notably, the lesion did not enhance with contrast. This resulted in mild cervicomedullary parenchyma compression but without significant edema, consistent with chronicity. The differential diagnosis for this lesion included: ependymoma, glioma, metastasis, primitive neuroectodermal tumor, or teratoma. Additional evaluation of the entire neuraxis with CT/MR imaging (MRI) documented a type I diastematomyelia from T10 to L4 level, with an osseous bar at L1/L2 [Figures 3 and 4].

Surgery

A suboccipital craniectomy provided exposure of the cranial/CMJ allowing for the identification of a plane to expose the tumor [Video 1]. The posterior arch of C1 was removed and widened bilaterally. Following a midline durotomy, a glistening white, fibrous capsule was identified surrounding the lesion that extended laterally into the C1 neural foramen where it was adherent to the C1 nerve root. After the root and capsule were coagulated, sectioned, and sacrificed, the capsule was dissected off the dura. The ventral cyst was then intentionally opened; a milky fluid erupted from inside the cyst, following which the capsule and solid encircling mass were excised. Ultimately, the cyst wall adjacent to the medulla was cut sharply (i.e., avoiding devascularizing the medulla), and the dura was then closed primarily. The intraoperative frozen section identified a nerve root ganglia and nerve root segments (likely C1), small vessels, and collagenous tissue.

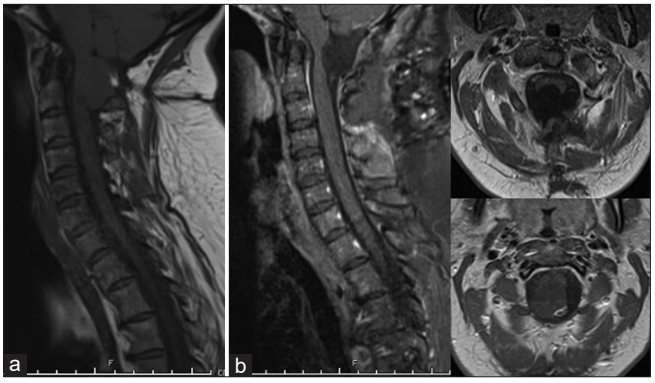


Figure 1: Preoperative sagittal cervical magnetic resonance imaging (MRI) T1-weighted sequence without contrast (a) and cervical MRI sagittal and two axial slices through the tumor T1-weighted sequence with contrast (b).

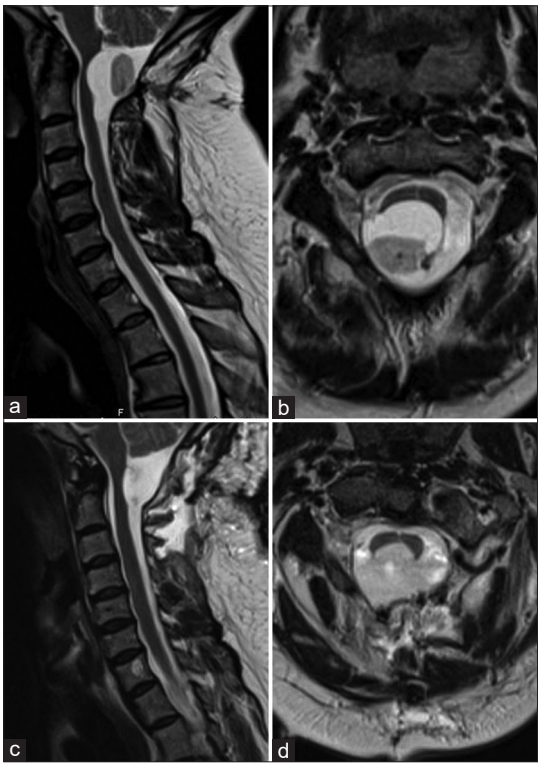


Figure 2: Preoperative cervical magnetic resonance imaging (MRI) T2-weighted sequence sagittal (a) and axial (b) views. Postoperative sagittal (c) and axial (d) views of a T2-weighted MRI of the cervical spine.

Pathological diagnosis

Histopathology of the solid lesion demonstrated a mature teratoma comprised of islands of cartilage, skeletal muscle, adipose tissue, respiratory epithelium, underlying mucous glands, and collagenous septae with rare psammoma bodies [Figure 5]. The cyst contents included scattered erythrocytes, lymphocytes, and cellular debris.

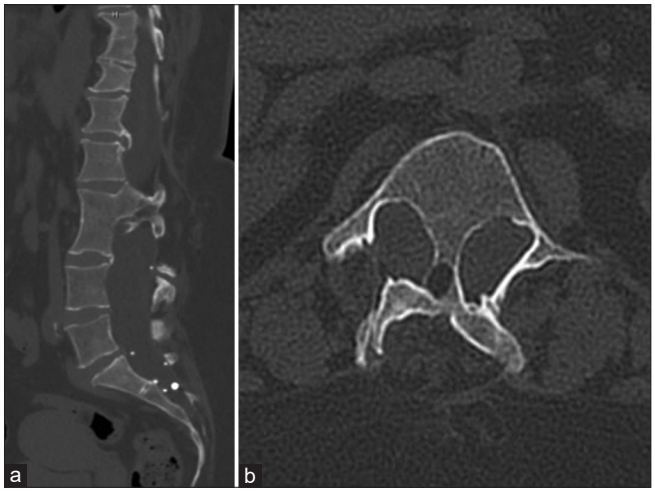


Figure 3: Computed tomography of the lumbar spine without contrast demonstrating the diastematomyelia and bony osseous bar at L1 to L2 levels (a: Sagittal view; b: Axial view).

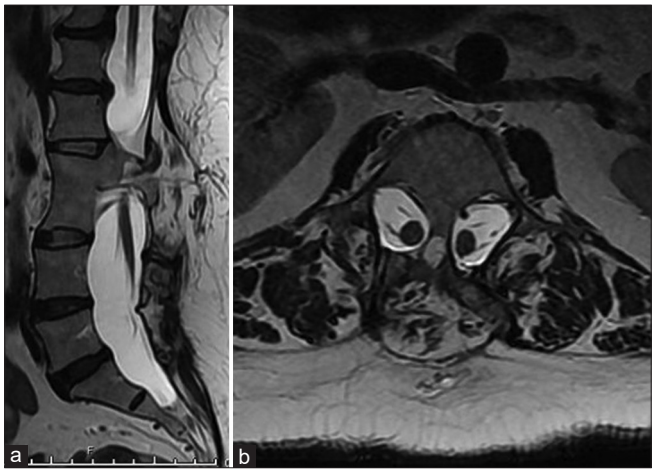


Figure 4: Magnetic resonance imaging of the lumbar spine T2-weighted sequence demonstrating the type I split cord malformation (a: Sagittal view; b: Axial view).

Postoperative outcome and follow-up

The immediate postoperative brain MRI demonstrated significant decompression of the central canal at the C2 level with subtotal resection of the mass [Figure 2]. By the fifth postoperative day, the patient was ambulating without assistance, and her left hemiparesis had improved to a 4+/5 level. She was discharged home. Two weeks later, the patient reported no further occipital headaches, and her left hemiparesis had fully resolved.

DISCUSSION

In the pediatric population, intradural teratomas are often associated with a variety of spinal anomalies at the same site, particularly dysraphism. However, in adults, they are most often isolated findings.[5,7,14] The literature was reviewed from 1928 to the present date and the relevant cases of intradural teratomas in adults with concurrent spinal dysraphism are shown in Table 1.[7] Notably, our patient had undergone posterior cervical resection of a C2 "mass" at the age of 14 and also demonstrated an incidental type I split cord thoracolumbar malformation. Makary *et al.* described an intramedullary mature teratoma at C1–2 with associated spinal dysraphism in an adult.[8] Bouaziz *et al.* reported a cervical intradural extramedullary teratoma in an adult without associated dysraphism or a history of prior spinal surgery.[4] There were two cases in India of children with cervical and thoracic intradural teratomas with an associated type II and type I split cord malformation, respectively, at the site of their lesions.[9,14] Notably, our case demonstrated mature pulmonary structures (mucous glands, collagenous septae, and psammoma bodies) on pathological analysis. Ramdial *et al.*, in 1998, also reported pulmonary differentiation and associated spinal dysraphism.[13]

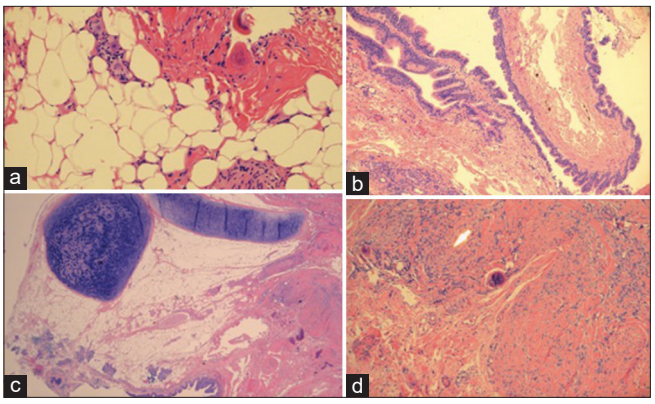
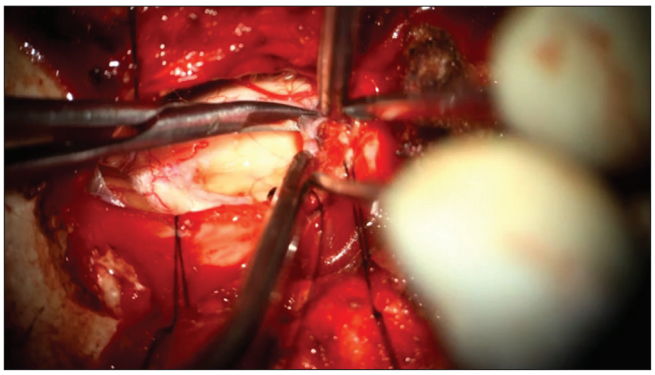


Figure 5: Frozen section with nerve root ganglia, nerve segments, small vessels, and collagenous tissue (a) and H&E permanent block demonstrating ciliated squamous epithelium (b), cartilage and respiratory mucosal tissue (c), nerve (arrow), and muscle tissue (d).



Video 1: Intraoperative video of microsurgical dissection and resection of a mature teratoma in the cervicomedullary junction.

Table 1: Summary of reported cases of intradural teratomas in adults with concurrent spinal dysraphism from 1928 to the present day.

First author(s), year	No. of cases	Gender	Age (years)	Location	Spinal Dysraphism	Prior Spine Surgery	Prior Trauma	Resection
Hosoi, 1931[6]	1	M	24	L2-L3	L5-S1 spina bifida	-	-	Incomplete
Bakay, 1956[3]	1	F	65	L1-L2	L1 and L2 vertebral body fusion	-	-	Incomplete
Pelissou-Guyotat *et al.*, 1988[12]	1	M	33	L4	L4 spina bifida occulta	No	No	Complete
Nicoletti *et al.*, 1994[10]	1	M	47	Conus medullaris	Conus medullaris caudal exophy	No	No	Incomplete
Ak *et al.*, 2006[1]	1	F	43	C2-C3	C3 spina bifida	No	No	Complete
Makary *et al.*, 2007[8]	1	F	46	C1-C2	C1-2 dysraphic congenital spinal malformation	No	No	Complete
Present case	1	F	52	Cranio-cervical junction	Diastematomyelia, Type I	Yes	No	Incomplete

M: Male, F: Female

Surgery

Surgical excision of symptomatic lesions remains the first line of treatment, although the recurrence rate of mature spinal teratomas is low, with comparable recurrence rates for gross total (i.e., 9–10%) versus subtotal resection (i.e., 10–11%).[2,5,7] Alternatively, Wan *et al.* reported that adults with intradural teratomas undergoing subtotal resection had 50% recurrence rates at 80 postoperative months versus no recurrences following gross total resection.[15] In addition, Prasad and Divya determined that recurrence rates were higher after subtotal resection, but observed that gross total resection could only be achieved in 45% of cases due to contiguous critical structures.[11]

Lack of efficacy of adjunctive therapies for teratomas

Due to the slow-growing nature of mature teratomas, radiotherapy is typically not considered unless there is a concern for malignancy on histology, while the efficacy of chemotherapy has not been explored.[2] In the present case, a subtotal resection was performed without adjunctive therapy. Given the indolent nature of these tumors and the risk of recurrence associated with subtotal resections, this patient required clinical and radiological long-term follow-up.

CONCLUSION

Intraspinal teratomas in adults are exceedingly rare. Here, we report a 52-year-old Hispanic female who at age 14 had undergone posterior C2 cervical surgery for a "mass," and now presented with a cervicomedullary intradural extramedullary mature teratoma that was successfully partially resected.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of Artificial Intelligence (AI)-Assisted Technology for assisting in the writing or editing of the manuscript and no images were manipulated using the AI.

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