Symptomatic mature teratoma of the lumbar spine: A case report

Coby Cunningham¹, Chiara Flores¹, Rocco Dabecco², Palgun Nisarga³, Janice Ahn³, Richard Williamson²

¹Department of Neurosurgery, Drexel University College of Medicine, Philadelphia, Departments of ²Neurosurgery and ³Pathology, Allegheny General Hospital, Pittsburgh, Pennsylvania, United States.

E-mail: *Coby Cunningham - cjc425@drexel.edu; Chiara Flores - caf346@drexel.edu; Rocco Dabecco - rocco.dabecco@ahn.org; Palgun Nisarga - palgun.nisarga@ahn.org; Janice Ahn - janice.ahn@ahn.org; Richard Williamson - richard.williamson@ahn.org

INTRODUCTION

Mature teratomas are one of the three WHO defined variations of teratoma; mature, immature, and teratoma with malignant transformation. Each is derived from two or more of the three embryonic layers: the endoderm, mesoderm, and ectoderm. Mature teratomas are the most well-differentiated variation, typically showing varying histologic morphologies (i.e., skin, hair, teeth, nerve, brain, smooth muscle, adipose tissue, cartilage, and enteric- or respiratory-type tissue). Further, they may demonstrate multicystic components that may be watery, mucoid, and/or sebaceous in consistency.
Mature teratomas, though variable in presentation, are classically found in young females. Only very rarely (0.2%) do they arise within the spinal cord (i.e., intradural/intradural/ extramedullary) at the cervical, thoracic, and lumbar levels. Here, we encountered a 78-year-old female with a subacute presentation of paraparesis attributed to an MR-documented intramedullary conus lesion with exophytic extension into the L1-L4 conus/cauda equina. Following excision of the teratoma, the patient largely regained normal neurological function.

**CASE DESCRIPTION**

A 78-year-old female with a history of a “spinal mass” first documented in 2014, presented with the subacute onset (i.e., over several weeks) of a bilateral paraparesis right greater than left [Table 1].

The lumbar MRI revealed an 81 × 30 × 25 mm intrathecal intramedullary conus mass with extramedullary extension into the lumbar canal at L1-L4. The mass was accompanied by multiple complex cysts [Figures 1a and b].

**Surgery**

The patient underwent an urgent L1-L4 laminectomy with removal of the conus-cauda equina multicystic tumor (i.e., a large and multilobulated cystic lesion compressing the neural elements). Fenestration of a cystic lobule resulted in extrusion of a nonpurulent milky fluid that proved to be sterile. The cyst was safely dissected off the conus and filum terminale with the use of intraoperative neuromonitoring. Interestingly, there was no gross evidence of accompanying teeth, hair, or visible adipose tissue. There were no intraoperative complications.

**Table 1:** Compilation of the relevant pre- and post-operative data for this case.

| Variable                        | Finding                  |
|---------------------------------|--------------------------|
| Variable                        | Finding                  |
| Age                             | 78                       |
| Sex                             | Female                   |
| Preoperative neurological examination |                          |
| Iliopsoas                       | Right 2/5                |
| Quadriceps                      | Left 4/5                 |
| EHL                             | Right 1/5                |
| DF                              | Left 5/5                 |
| PF                              | Right 3/5                |
| Sensory deficit                 | Left 5/5                 |
| MR finding                      | Bilaterally intact       |
| Immediate postoperative neurological examination | 81×30×25 mm intrathecal intramedullary multicystic mass at L1–L4 involving the conus medullaris and cauda equina |
| Iliopsoas                       | Right 5/5                |
| Quadriceps                      | Left 5/5                 |
| EHL                             | Right 5/5                |
| DF                              | Left 5/5                 |
| PF                              | Right 5/5                |
| Pathology                       | Left 5/5                 |
| A                               | Multiphasic: solid and cystic components. All components were mature |
| B                               | (Endoderm): pseudostratified ciliated and nonciliated columnar epithelium with focal stratified squamous epithelium |
| C                               | (Mesoderm): adipose tissue, hyaline cartilage, and smooth muscle bundles |
| D                               | (Ectoderm): peripheral nerve bundles and Pacinian corpuscles |

PF: Plantar flexors, DF: Dorsiflexors, EHL: Extensor hallucis longus
Postoperative course and imaging

Immediately postoperatively, the patient was neurologically intact. However, on postoperative day 2, she experienced an acute change in neurologic status of unknown etiology (i.e., severe disorientation, persistent gaze preference with facial droop, and left-sided hemiparesis). CT brain was immediately conducted following the acute decline and showed no evidence of a potentially causal pathology. Subsequently, MR brain and EEG were performed to definitively rule out a new stroke or lesion and seizure activity, respectively. Both were unremarkable. The patient was eventually discharged fully orientated and with progressively improving lower extremity strength. Imaging obtained on postoperative day 0 showed successful resection of the mass without evidence of acute complication [Figure 2].

Histology of mature teratoma

Microscopically, the tumor was composed of both solid and cystic components (i.e., that contained blood/debris and showed predominantly pseudostratified ciliated and nonciliated columnar epithelial lining with focal stratified squamous epithelium [endoderm]). The surrounding fibrous connective tissue contained lobules of mature adipose tissue (mesoderm), mature hyaline cartilage (mesoderm), peripheral nerve bundles (ectoderm), Pacinian corpuscles (ectoderm), and focal smooth muscle bundles (mesoderm) [Figures 3 and 4]. No immature structures were seen.

DISCUSSION

Classification of teratomas

Historically, cystic intraspinal lesions including teratomatous cysts, neurenteric cysts, and teratoid cysts have demonstrated varying morphologic criteria. Here, a definitive diagnosis of a mature cystic teratoma was established in accordance with the current WHO classification guidelines (i.e., tissues originating from all three embryonic cell layers were clearly demonstrated within the presence of only highly differentiated cells).
CT/MR diagnoses for intramedullary/extramedullary conus/cauda equina mature teratoma

The differential diagnosis for an intradural intramedullary/extramedullary/exophytic conus-cauda equina spinal mass includes myxopapillary ependymoma, paraganglioma, nerve sheath tumor, meningioma, and rarely spinal teratomas.[5] For mature spinal teratomas, CT scans may confirm the “sack-of-marbles” appearance due to fat globules within the cystic portion of the mass (93% of cases) and calcification due to the presence of teeth (53% of cases).[6] CT may also demonstrate an intracystic mass adherent to the wall of the teratoma, known as a Rokitansky nodule (hair). On T1-weighted MR images, these lesions may be hyperintense due to their intrinsic fat and high fluid content.[8] Although imaging findings are often highly suggestive of mature teratomas, definitive diagnosis requires surgical resection and histologic examination of the tissue/mass.

Pathogenesis of mature spinal teratomas

The pathogenesis of spinal teratoma still remains unclear; however, theories include (1) tumors arising from misplaced primordial germ cells from the yolk sac, (2) tumors arising from pluripotent cell rest at sites of neural closure, and (3) malformations giving rise to a dysembrogenic process. Mature teratoma has also been associated with spinal dysraphism, lumbar puncture, and prior spinal surgery.[6,9] Notably, mature teratomas within the spinal cord are slow growing, with an average growth of 1.8 mm/year.[8]

Mature teratomas: Risk of malignant transformation

Mature teratomas have a 2% risk of malignant transformation and a <1% risk of cyst rupture.[7,4] Thus, diagnosis of a mature teratoma on MR/CT imaging warrants timely surgical intervention regardless of neurological deficits to avoid these potentially severe sequelae.

Efficacy of surgical resection of mature teratomas

As in this case, surgical removal of mature teratomas typically results in significant neurological improvement.[1] As early diagnosis and gross total resection are correlated with the best surgical outcomes, early diagnostic evaluation and treatment are warranted.

CONCLUSION

Intrathecal intramedullary/extramedullary mature teratomas of the conus that results in subacute cauda equina syndromes are rare. The differentiate diagnosis of such lesions is exceptionally broad, but with rapid identification, these mature teratomas may be readily and completely resected. As was seen here, these patients may, more often than not, go on to extremely favorable neurologic recovery.

Declaration of patient consent

Patient’s consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Fetcko K, Dey M. Primary central nervous system germ cell tumors: A review and update. Med Res Arch 2018;6:1719.
2. Hattab EM. Germ cell tumors. In: Arie Perry DJ, editor. Practical Surgical Neuropathology: A Diagnostic Approach. 2nd ed. Amsterdam, Netherlands: Elsevier; 2018.
3. Kawai M, Nagoshi N, Iwanami A, Mikami S, Tsuji O, Fujita N, et al. Acute regrowth and dissemination of a mature spinal cord teratoma after partial resection. BMJ Case Rep 2018;2018:bcr201723742.
4. Kleihues P, Louis DN, Scheithauer BW, Rorke LB, Reifenberger G, Burger PC, et al. The WHO classification of tumors of the nervous system. J Neuropathol Exp Neurol 2002;61:215-25; discussion 226-19.
5. Koeller KK, Shih RY. Intradural extramedullary spinal neoplasms: Radiologic-pathologic correlation. Radiographics 2019;39:468-90.
6. Koen JL, McLendon RE, George TM. Intradural spinal teratoma: Evidence for a dysembryogenic origin. Report of four cases. J Neurosurg 1998;89:844-51.
7. Mandal S, Badhe BA. Malignant transformation in a mature teratoma with metastatic deposits in the omentum: A case report. Case Rep Pathol 2012;2012:568062.
8. Outwater EK, Siegelman ES, Hunt JL. Ovarian teratomas: Tumor types and imaging characteristics. Radiographics 2001;21:475-90.
9. Pandey S, Sharma V, Shinde N, Ghosh A. Spinal intradural extramedullary mature cystic teratoma in an adult: A rare tumor with review of literature. Asian J Neurosurg 2015;10:133-7.
10. Sung KS, Sung SK, Choi HJ, Song YJ. Spinal intradural extramedullary mature cystic teratoma in an adult. J Korean Neurosurg Soc 2008;44:334-7.

How to cite this article: Cunningham C, Flores C, Dabecco R, Nisarga P, Ahn J, Williamson R. Symptomatic mature teratoma of the lumbar spine: A case report. Surg Neurol Int 2022;13:16.