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Intradural Cavernous Lymphangioma of the Thoracic Spine: Case Report, Technical Considerations, and Review of the Literature

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Abstract

Background Context
Cavernous lymphangioma is a rare slow growing tumor that can cause neurologic compromise when it involves the central nervous system. Involvement of the spinal column is rare but may involve the osseous structures or the epidural space of the spinal column.

Purpose
We report the first case of an intradural, extramedullary cavernous lymphangioma involving the thoracic spinal cord.

Methods
An eighty-three year old female presented with progressive gait ataxia, bilateral lower extremity weakness and a band-like sensation in the middle and lower thoracic dermatomes. Magnetic resonance imaging of the thoracic spinal cord revealed hyperintensity on T2 and enhancement of an intradural cystic mass along the dorsal aspect of the T5-8 levels with significant compression of the spinal cord.

Results
Complete surgical resection was difficult due to the adherence of the tumor to the pial surface and microvasculature of the thoracic spinal cord. Recurrence of the mass was ultimately treated with cystic fluid diversion into the peritoneum. At her twenty-eight month follow-up visit, the patient was able to ambulate with minimal assistance. A comparative literature review is presented. There are no reports in the literature of intradural thoracic spinal cord involvement.

Conclusions
Intradural cavernous lymphangioma of the spine poses a unique surgical challenge for complete resection. Cystic fluid diversion appears to be a viable treatment option with lasting benefit if complete resection is not achieved.

Introduction

Lymphangiomas are benign, slow-growing soft tissue tumors with predilection for the head and neck but may involve any organ. Lymphangiomas are classified into congenital or acquired forms and histologically divided into cavernous, capillary or cystic type. [1,8,11,12,16,17,19–21] Tumor involvement of the spinal column is exceedingly rare and presents either as a primary osseous tumor or as a soft tissue tumor in the epidural space.[5,10,11,13,16,18,20] When lymphangiomas involve the epidural space, it usually occur as an isolated mass or an extension from a primary mediastinal lesion. Treatment for symptomatic lymphangiomas of the spine requires complete surgical resection. Recurrence is common when residual disease is left behind.[5,8,11,17,19] We report the first case of an intradural, extramedullary cavernous lymphangioma (CL) involving the thoracic spinal cord.

Clinical Case Report

An eighty-three year old right-handed female with a history of shingles presented to the clinic with an eight-month history of an abnormal band-like sensation in the middle and lower thoracic dermatomes and progressive gait ataxia requiring a walker for ambulation. Physical examination was significant for 5/5 bilateral lower extremity strength, decreased proprioception in the left lower extremity, positive Romberg Sign, decreased sensation over the mid-thoracic dermatomes, and 3+ bilateral patellar and Achilles tendon reflexes.

Initial magnetic resonance imaging (MRI) of the thoracic spine revealed an intradural, extramedullary cystic lesion along the dorsal aspect of T5-8 levels with significant compression of the thoracic cord (Fig. 1A-B). Systemic imaging included a PET scan and CT of the chest, abdomen and pelvis which were unrevealing for a contributing disease process.

The patient underwent a T5-8 laminectomy and dural opening for lesion resection. Intraoperative neuro-monitoring included somatosensory evoked potentials, motor evoked potentials, and electromyographic modalities. During surgical exploration the cyst was found to be dark in

color with a thick and uniform fibrous capsule. The capsule was incised revealing opaque fluid with no septations. Under the microscope, sharp dissection was utilized for successful release of the dural surface of the tumor. However, attempts at dissecting the capsule from the pial surface proved problematic as the microvasculature of the pia and capsule were intimately involved. As such, residual capsule was left attached to the pial surface. (Fig. 1C-D). There were no electrophysiological changes noted intraoperatively. Final pathological review was consistent with cavernous lymphangioma. (Fig. 2A-B)

At her four week follow-up, the patient was able to walk independently but continued to experience decreased proprioception in the left lower extremity and hyperactive (3+) reflexes in both her patellar and Achilles tendons. However at four months, she presented with new onset weakness (3/5) in dorsiflexion of the left lower extremity, a positive Romberg sign, and progressive gait ataxia requiring a walker. Repeat MRI done at this time revealed accumulation of fluid within the cystic mass, now extending between T5-9 with compression of the thecal sac (Fig. 3A). The patient returned to surgery for another incomplete resection (Fig. 3B). Immediately after surgery, her dorsiflexion strength (5/5) and gait ataxia improved although she continued to require a walker for ambulation.

Three months later, she presented again with progressive neurological decline. Examination showed bilateral lower extremity strength of 4/5 with severe ataxia requiring a wheelchair for mobility. Repeat MRI revealed tumor recurrence with cord compression (Fig. 3C). Given the large cystic component of the mass, the patient was taken back to the operating room for placement of a cysto-peritoneal shunt. Her post-operative course was consistent with slow neurological improvement. At her nine month follow-up visit, lower extremity strength was 5/5 and she was ambulating with a walker. During her latest follow-up visit twenty-eight months after shunt placement, the patient's neurological examination plateaued and remained stable as she continued to ambulate with a walker and maintained 5/5 strength in her lower extremities.

Discussion

Lymphangiomas are characterized by abnormal dilated lymphatic channels and cystic spaces lined with endothelium of varying sizes containing lymphocytes.[4,8,19,19,22] These benign hamartomatous tumors are most commonly found within the first two years of life and often involve the head and neck.[1,19,20] Lymphangiomas are thought to be true congenital malformations of large ectatic and sequestered lymphatic channels that fail to connect with venous blood vessels or normal lymph tissue.[4,22] Histologically, lymphangiomas are classified into one of three variants: cystic, cavernous, or capillary lymphangiomas. Cavernous lymphangiomas are composed of thin walls and contain opaque or milky fluid within the lumen consistent with findings in this reported case.[5,8,20,22]

Lymphangioma involvement of the spinal column is very rare. When lymphangioma tumors involve the spinal column, it occurs either as a primary soft tissue tumor with epidural involvement, a primary osseous tumor, or a combination of both. The first reported case of primary lymphangioma involving the vertebral body of the spinal column was in 1956 by Falkmer and Tilling. [7,18] In 2002, Mendez *et al*. documented nine cases of primary lymphangioma involving the bone, only five of which involved the spinal column.[13,18] Involvement of the osseous structures of the spinal column may result in devastating outcomes as massive osteolysis takes place, resulting in severe destruction and instability of the spinal cord, especially at the craniocervical junction.[3,6]

The first reported case of a primary soft tissue lymphangioma involving the epidural space was described by Saito *et al.* in 1999 presenting as a dumbbell tumor.[20] Several case reports followed describing epidural spinal cord involvement in which the tumor originated from neural origin with subsequent extra-spinal involvement.[5,10,11,16] Epidural spinal cord involvement may also be due to invasion of a mediastinal mass into the intervertebral foramen, subsequently involving the

nerve root and epidural space.[17] However, only one prior case of an intradural lymphangioma tumor involving the sacral thecal sac has been reported.[12]

As the central nervous system is devoid of lymphatic tissue, lymphangiomas are thought to spare the intradural space and when identified, the pathophysiology remains controversial.[11,21] Several possible explanations have been proposed. The first involves the lymphatic tissue within the intervertebral foraminal fat pad. It is possible that remnant tissue is left within the thecal sac during development.[4,14,20] Another plausible explanation may be the involvement of lymphatic channels as they travel along neurovascular bundles, which is important for cerebrospinal fluid drainage through small arachnoid processes penetrating the dura. This could hypothetically provide a conduit for introduction of lymphatic tissue into the intradural space.[4,11,14,20] Acquired lesions may be due to obstruction of lymphatic flow and subsequent enlargement of lymphatic tissue.[5,10,21] Finally, it is thought that the periosteum of bone has a vast plexus of lymphatic capillaries which can penetrate the intradural space iatrogenically or with trauma.[5,7,20]

Radiographic diagnosis of a lymphangioma tumor of the spinal column remains challenging given its nonspecific characteristics. It is established that MRI has superiority over computed tomography (CT) because of the ability to define the boundary, size, and loculations if present. Our imaging findings remain consistent with the few case reports of cavernous lymphangioma involving the spinal column: low to isointense on T1, high intensity on T2, and homogenous cyst enhancement with intravenous gadolinium on MRI examination. However, whether cavernous lymphangiomas involve the intradural or epidural space remains difficult to definitively identify on preoperative imaging alone. Differential diagnosis for an intradural thoracic cystic mass is extensive and includes metastatic tumor, schwannoma, meningioma, neurofibroma, sarcoma, intraspinal arachnoid cyst, lipoma, embryonal tumor, hemangioma, meningothelioma, and other cystic structures.[5,10,11,20]

Cavernous lymphangioma presents with neurologic decline due to its mass effect. Complete excision is the primary goal for successful treatment as reviewed in the literature (Table 1). [1,19,22] However, as noted in this report, total resection may not be possible. Aggressive attempts at developing tissue planes or dividing shared vascularity of tumor with pial structures can result in an ischemic cord event. Recurrence of lymphangioma is due to incomplete resection rather than malignant transformation.[8,17] Prior resection, trauma, infection and hemorrhage of a primary soft tissue lymphangioma may precipitate an "aggravated tumor" resulting in worsening mass effect and subsequent neurologic decline. This is a well-documented phenomena in other organs of the body where lymphangioma occurs more commonly.[1,11,16,17,19,21] Sole treatment options in peripheral organs include surgical resection and possible adjuvant sclerotherapy as it is not sensitive to radiation or chemotherapy.[1,11,19]

As reported in this case, the intradural capsule of the tumor was adherent to the microvasculature and pia of the spinal cord, thus impeding a complete resection. Although residual tumor remained, removal of the mass effect allowed neurologic symptoms to temporarily improve. Since surgical debulking of the tumor provided symptomatic management, a permanent method of decompression was attempted by placement of a cysto-peritoneal shunt. Although the pathophysiology of intradural lymphangiomas is not well understood, clinical stability after shunting suggests that fluid diversion may aid in symptomatic treatment.

Cystoperitoneal shunt placement is an established treatment for developmental and traumatic symptomatic syringomyelia with reasonable clinical outcomes reported in the literature.[2,9,15] Longer follow-up is likely needed to evaluate the longevity of this treatment for recurrent CL of the spinal cord. It is possible that the viscous cystic fluid seen in cavernous lymphangiomas may lead to shunt obstruction and hence recurrence of symptoms.

Conclusion

We describe the first case of an intradural, extramedullary cavernous lymphangioma of the thoracic spine with intradural involvement. Caution is advised with attempts of complete resection given the adherence of the tumor capsule to the microvasculature and pia. Consideration should be given to a cystic fluid diversion procedure if complete surgical removal is not possible.

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Figures and Table Captions

Figure 1.

Pre- and post-operative MRI of the thoracic spine. T2 weighted pre-operative (A) sagittal and (B) axial images demonstrating a cystic intradural mass at the T5-T8 levels with cord compression. Post-operative (C) sagittal and (D) axial images demonstrating a right side laminectomy and durotomy with partial decompression of the spinal cord.

Figure 2.

Hematoxylin and Eosin (H&E) stain (40x) cross section of lesion. (A) Thin-walled fibrous tissue with lymphatic channels lined by flat endothelial cells and (B) dilated blood vessels filled with red blood cells, consistent with CL.

Figure 3.

T2 weighted MRI's of the thoracic spine. (A) Sagittal image demonstrating recurrence of CL along the T5-T9 intradural space. (B) Sagittal post-reoperative image showing decompression of the thoracic cord. (C) Sagittal MRI demonstrating re-recurrence of the CL after second surgery.

Table 1

Review of reported cases of primary soft tissue lymphangiomas involving the spine.

Author	Year	Age	Gender	Spine Location	Dural Location	Pathology (Lymphangioma)	Treatment	Postoperative follow-up time (months)	Postoperative neurologic function
Saito[20]	1996	56	F	Lumbar	Extradural	Cavernous	Lami L4	23	Improved
Kanamori[12]	2002	56	M	Sacral	Intradural	Cystic	Lami S2-S3	66	Complete recovery
Jiang[11]	2005	47	M	Sacral	Extradural	Cystic	Lami S2-S3**	12	Complete recovery
Jiang[11]	2005	12	M	Lumbar	Extradural	Spongiform*	N/A	12	Complete recovery
Ha[10]	2005	16	M	Thoracic	Extradural	Cystic	Lami T5-T7	4	Complete recovery
Chu[5]	2007	61	F	Cervical/Thoracic	Extradural	Cavernous	Lami C6-T1	12	Improved
McLoughlin[17]	2008	1	M	Cervical/Thoracic	Extradural	Cavernous	Lami C5-T2	48	Improved
Lee[16]	2011	43	F	Cervical	Extradural	Cavernous	Hemilami C6-C7	12	Complete recovery

*Not typical classification of lymphangioma soft tissue tumors
**Surgery based on imaging in case report
Key: F-Female, M-Male, N/A-not available, Lami-laminectomy, L- Lumbar, T-Thoracic, C-Cervical, S-Sacral