IJMS
Vol 44, No 2, March 2019

Case Report

Lymphangioma of the Thoracic Spine with Epidural Compression: A Case Report

Arash Fattahi[1], MD; Morteza Taheri[2], MD; Mina Majdi[3], MD

[1]Department of Neurosurgery, 7tir Hospital, Iran University of Medical Sciences, Tehran, Iran;
[2]Department of Neurosurgery, Rasool Akram Hospital, Iran University of Medical Sciences, Tehran, Iran;
[3]Department of Pathology, Gerash Hospital, Gerash University of Medical Sciences, Gerash, Iran

Correspondence:
Morteza Taheri, MD;
Department of Neurosurgery, Rasool Akram Hospital, Postal code: 14456-13131, Tehran, Iran
Tel: +98 912 0194908
Fax: +98 21 66509120
Email: drtaheri38@yahoo.com
Received: 12 December 2017
Revised: 8 April 2018
Accepted: 15 April 2018

Abstract

Lymphangiomas are benign lesions consisting of abnormal proliferations of lymphatic vessels. Lymphangiomas associated with bone involvement, particularly in vertebral bodies, accompanied by cord compression, are extremely rare, and our literature review yielded only a few relevant reports. We describe a 61-year-old man presenting with progressive paraparesis and sphincter disturbance of 5 months' duration. Magnetic resonance imaging (MRI) revealed an enhancing T8 vertebral body involvement as well as a homogeneously enhancing posterior epidural mass at the T7–T8 level, with severe cord compression and cerebrospinal fluid (CSF) blockade. The patient underwent surgery via T7–T8 laminectomy, and after the removal of the epidural mass, the surgical procedure was stopped due to severe bleeding. Histopathologic examination reported a lymphangioma. After 10 days, the patient was able to walk. In the sixth postoperative month, MRI showed complete relief of the mass effect. The wide spectrum of the preoperative differential diagnosis of lymphangiomas renders a definite preoperative diagnosis impossible; therefore, histopathologic examination is the sole definite route for their diagnosis. In case of the solitary lymphangiomas of the spine with epidural compression, preoperative angiography and embolization should be considered to reduce intraoperative bleeding and enhance the chance of total resection and total surgical resection should be performed to decrease the likelihood of recurrence.

Please cite this article as: Fattahi A, Taheri M, Majdi M. Lymphangioma of the Thoracic Spine with Epidural Compression: A Case Report. Iran J Med Sci. 2019;44(2):172-175. doi: 10.30476/IJMS.2019.44519.

Keywords ● Spine ● Lymphangioma ● Neurologic deficit ● Pathology

What's Known

• Lymphangiomas are benign lesions originating from lymphatic tissues due to the failure of lymphatic channels to communicate with normal lymphatic or venous systems. They consist of abnormal lymphatic channels covered by the endothelium. They are more prevalent in children than adults, with soft tissues being the most commonly involved sites.

What's New

• Solitary thoracic vertebral body lymphangiomas associated with epidural cord compression and progressive neurological deficits are extremely rare. Our case demonstrates that lymphangiomas should be in the differential diagnosis of multiloculated vertebral body lesions. Preoperative angiography/embolization should be considered to reduce intraoperative bleeding and enhance the chance of total resection.

Introduction

Lymphangiomas are benign lesions with abnormal proliferations of lymphatic vessels.[1] These lesions are usually found in soft tissues; therefore, solitary bone lymphangiomas, particularly in vertebral bodies, associated with epidural cord compression are extremely rare.[2] We describe a male patient with progressive myelopathy due to a thoracic spine lymphangioma and review his diagnosis and treatment strategy.

Case Presentation

A 61-year-old healthy man presented to our hospital in the Iranian city of Gerash in November 2015 with progressive paraparesis and sphincter disturbance (urinary urgency) of 5 months' duration. Physical examination showed paraparesia (muscle

strength=3/5 based on a 5-point scale of muscle strength score), hyper-reflexia, and a positive Babinski sign. The patient was unable to walk. Unfortunately, he had previously been treated with a diagnosis of lumbar canal stenosis. A whole-spine magnetic resonance imaging (MRI) with and without gadolinium (Gad) was performed, which demonstrated an enhancing T8 vertebral body lesion associated with a homogeneously enhancing posterior epidural mass at the thoracic spine (T7–T8 level), with severe cord compression and cerebrospinal fluid (CSF) blockade (figure 1).

Due to neurological deficits and cord compression on imaging, surgical decompression via the posterior approach was performed. On prone position, following total laminectomy of T7 and T8 and superior hemilaminectomy of T9, severe bleeding occurred from the left pedicle of the T8 vertebral body and the epidural mass. The bone had multiple cavities, causing severe venous bleeding, associated with a reddish-brown soft suckable epidural mass. As much as possible, the pedicular and epidural mass was resected from a lateral route. Unfortunately, due to severe bleeding (approximately 3.5 lit of blood loss), tumor removal was discontinued. Pedicle-screw fixation, followed by arthrodesis from T6 to T10, was performed. Postoperative computed tomography (CT) revealed an intact T8 vertebral body, and the complete removal of the posteriorly involved bony elements (figures 2a and 2b). Histopathologic examination revealed a lymphangioma for the specimens, involving bone and extradural mass (figures 2c and 2d).

On the 10th postoperative day, the patient was able to walk and after 6 months, he completely recovered. Six months after surgery, follow-up MRI showed complete relief of the mass effect without any epidural mass (figure 3). We decided to follow up the patient for a longer time for a probable subsequent clinical or imaging recurrence.

Informed consent was obtained from the patient for the publication of this case.

Discussion

Lymphangiomas are congenital benign lesions originating from lymphatic tissues due to the failure of lymphatic channels to communicate with normal lymphatic or venous systems.[1]

They are characterized by abnormal dilated lymphatic channels and cystic spaces covered by endothelium.[1] They are more prevalent in children than adults, with soft tissues being the most commonly involved sites.[2] Our literature review yielded only a few cases with the solitary lymphangiomas of the vertebral body accompanied by epidural cord compression.[1, 2-4]

Based on imaging studies, lymphangiomas have no distinctive appearance on CT scan; nonetheless, they can sometimes present with the coarse trabeculation of the vertebra while the shape of the vertebra is preserved.[5, 6]

On MRI, lymphangiomas have a low (due to serum) to intermediate (due to sedimentary packed cells) signal intensity on T1 and low (due to high cellular content) to high (due to high fluid content) signal on T2, which can be enhanced after Gad injection.[4] Our patient had the same preoperative imaging features.

The most important differential diagnoses of lymphangiomas include hemangiomas,

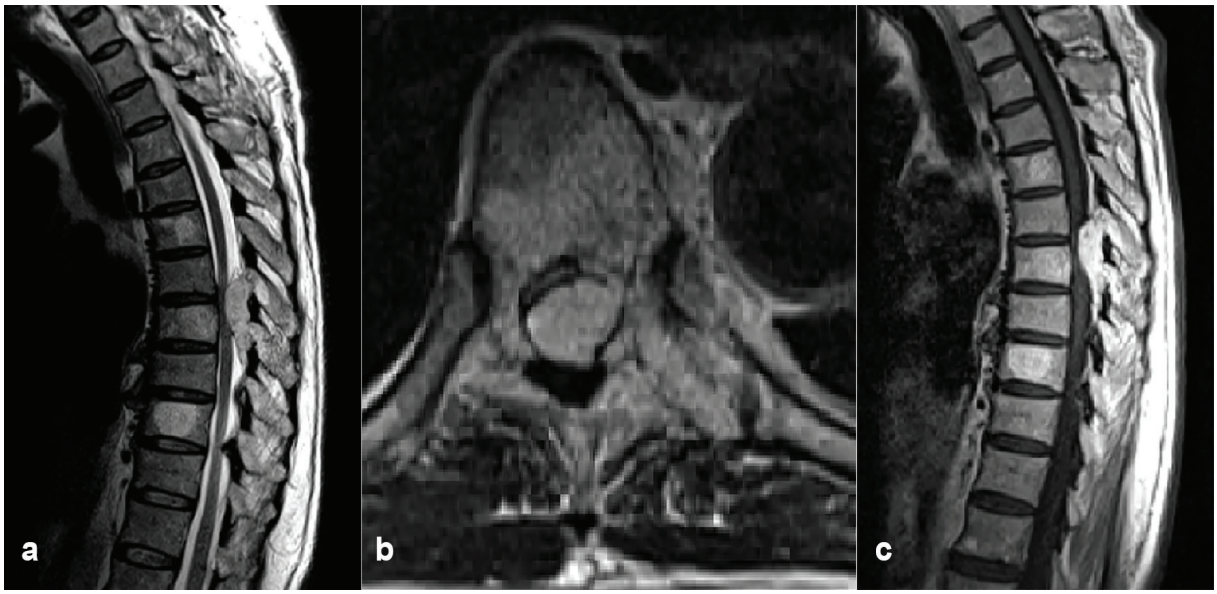

Figure 1: Magnetic resonance imaging features of the presented case shows an extradural extramedullary mass in the posterior epidural space compressing the spinal cord a) sagittal T2, b) axial T1, and c) axial T1 with Gad injection.

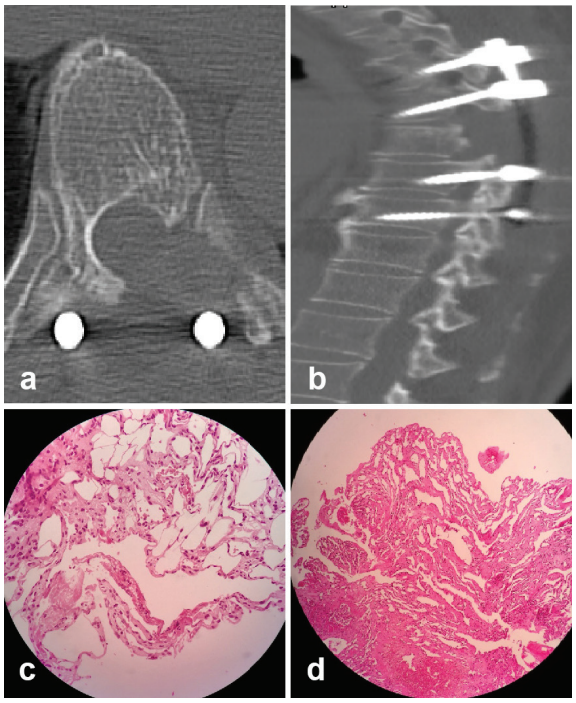

Figure 2: Postoperative computed tomography scan suggests laminectomy and spinal fixation a) axial and b) sagittal and c and d) demonstrates the histopathology features of the lymphangioma. The histopathologic specimen was stained with hematoxylin and eosin (H&E), and the microscopic images were taken with an Olympus C×31 microscope (×100 and ×400 power).

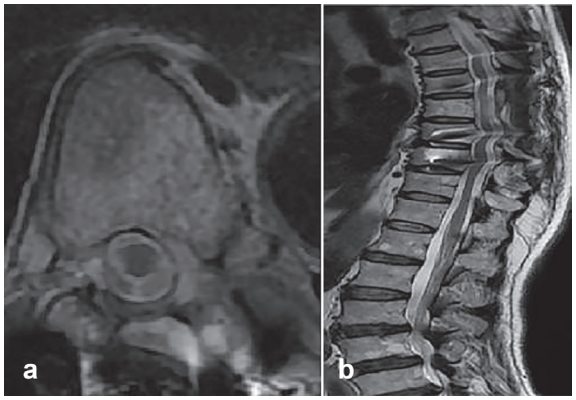

Figure 3: Magnetic resonance imaging a) axial T2 and b) sagittal T2 of the patient at 6 months' postoperative follow-up suggests no spinal tumor recurrence.

intraspinal arachnoid cysts, neurilemomas, meningotheliomas, lipomas, embryonal tumors, enterogenous cysts, metastatic tumors, and other rare intraspinal tumors such as hemangiopericytomas.[6] This wide spectrum of possible preoperative diagnosis renders the preoperative diagnosis of lymphangiomas, solely based on imaging characteristics, impossible and histopathologic examination is the only definite approach for their proper diagnosis.[6]

Histopathologic evaluations demonstrate multiple interconnecting lymphatic channels covered by single-layer endothelial cells where these cavities are filled with high protein materials.[4] Histopathology classifies lymphangiomas into three types: cystic, cavernous, and capillary.[4] The pathologic finding of our case was the same as this explanation.

Complete surgical resection is the best curative treatment option indicated in cases of spinal instability or neurological deficit.[2] In our patient, severe intraoperative bleeding resulted in an incomplete surgical removal of the lesion. We maintain that preoperative selective angiographic embolization enabled us to manage this case better; nevertheless, it entails that lymphangioma be at the top of the list of preoperative differential diagnoses.

Other alternative treatment options such as radiation, chemotherapy, embolization, aspiration, sclerotherapy, and recombinant interferon are reserved for inoperable cases, and they can be chosen according to clinical situations.[2]

Despite the benign nature of lymphangiomas, local tumor recurrence is possible.[2] Probable causes of recurrence are incomplete surgical resection, aspiration, and sclerotherapy.[2, 7] Accordingly, long clinical and radiological follow-up is mandatory, particularly in patients with incomplete resection.

Chu and others[2] described a 61-year-old woman presenting with an extradural lymphangioma at the C6–T1 level, which showed iso-signal intensity on T1 and high signal on T2 together with inhomogeneous enhancement after Gad injection. After resection, histopathology examination confirmed a cavernous lymphangioma. No recurrence occurred until 1 year's follow-up.

Jiang and others[6] reported 2 cases with a 1-year follow-up period. The first was a 47-year-old man presenting with an intradural cystic lymphangioma at the S2–S3 level, which demonstrated low-signal and high-signal enhancement on T1 and T2 and homogeneous enhancement on Gad injection. The second case was a 12-year-old male patient presenting with an extradural spongiform-type lymphangioma at the L1–L5 level, which demonstrated an iso- and high-signal mass on T1 and T2, respectively, and irregular enhancement after Gad injection.

Ha and colleagues[3] revealed an extradural lymphangioma (CD34 and D2-40 positive), associated with significant enhancement along its membrane, in a 16-year-old male patient at the level of T5–T7.

Kerolus and others[1] described an 83-year-old woman with an intradural extramedullary cavernous lymphangioma at the level of T5–T8, which demonstrated low- to iso-signal on T1 and

high signal on T2 as well as homogenous cyst enhancement on Gad injection. Four months after resection, she experienced recurrence.

McLoughlin and colleagues[7] reported a 1-year-old male patient with a mediastinal lymphangioma, which had invaded the extradural space at the C5–T2 level. He was treated via thoracotomy/laminectomy.

Aslan and others[8] described a 32-year-old man with a lymphangioma characterized by low signal on T1, high signal on T2, and faint wall and septal enhancement on Gad injection.

Conclusion

Our patient presented with an extremely rare case of a solitary thoracic vertebral body lymphangioma, resulting in progressive neurological deficits due to epidural cord compression. It demonstrates that lymphangiomas should be in the differential diagnosis of multiloculated vertebral body lesions. Preoperative angiography and embolization should be considered so as to reduce intraoperative bleeding and enhance the chance of total resection, and total surgical resection should be performed to decrease the likelihood of recurrence.

Conflict of Interest: None declared.

References

1 Kerolus MG, Patil J, Kurian A, Sani S. Intradural cavernous lymphangioma of the thoracic spine: case report, technical considerations, and review of the literature. Spine J. 2016;16:e561-5. doi: 10.1016/j.spinee.2016.03.012. PubMed PMID: 26970599.

2 Chu M, Li G, Wei L, Lin Y, Qi J, Wang C, et al. A rare case of cavernous lymphangioma in the epidural space of the cervicothoracic spine. Spine (Phila Pa 1976). 2007;32:E48-51. doi: 10.1097/01.brs.0000250992.55556.fe. PubMed PMID: 17202881.

3 Ha BY, Park JB, Kim YM, Lyo IU. Lymphangioma in the epidural space of the thoracic spine. J Korean Neurosurg Soc. 2010;47:403-5. doi: 10.3340/jkns.2010.47.5.403. PubMed PMID: 20539805; PubMed Central PMCID: PMCPMC2883066.

4 Lee SC, Moon SG, Kim NR, Choe WJ, Moon WJ. Cervical epidural lymphangioma presenting as a hemorrhagic cyst: a case report. Spine (Phila Pa 1976). 2011;36:E1117-20. doi: 10.1097/BRS.0b013e3181ffe9a7. PubMed PMID: 21304431.

5 Patnaik S, Jyotsnarani Y, Uppin SG, Susarla R. Imaging features of primary tumors of the spine: A pictorial essay. Indian J Radiol Imaging. 2016;26:279-89. doi: 10.4103/0971-3026.184413. PubMed PMID: 27413280; PubMed Central PMCID: PMCPMC4931792.

6 Jiang YG, Xiang J, Zhang L. Intraspinal lymphangioma: 2 case reports and literature review. Surg Neurol. 2006;66:430-6. doi: 10.1016/j.surneu.2005.12.038. PubMed PMID: 17015133.

7 McLoughlin GS, Nuchtern JG, Dauser RC, Sciubba DM, Gokaslan ZL, Wolinsky JP. Mediastinal lymphangioma presenting as an acute epidural hematoma. J Neurosurg Pediatr. 2008;1:474-6. doi: 10.3171/PED/2008/1/6/474. PubMed PMID: 18518699.

8 Aslan K, Bekci T, Gunbey HP, Incesu L, Turgut E. A rare cause of backache: epidural lymphangioma of the cervicothoracic spine. Spine J. 2015;15:1900. doi: 10.1016/j.spinee.2015.04.022. PubMed PMID: 25912507.