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Lumbar Extradural, Intra- and Extraforaminal Cavernoma Causing Lumbar Pain: A Case Report and Review of the Literature

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Abstract

Introduction: A spinal, intra- and extraforaminal epidural cavernoma is a very rare entity and can be easily misdiagnosed radiologically. Spinal extradural cavernomas can cause mild to devastating permanent neurological symptoms and should be removed before irreversible deficits occur.

Case Presentation: We present an uncommon case of a 50-year-old woman with lower back pain and a hypoesthesia of the ventromedial area of the thigh of the left leg. MRI (T2-weighted images, T1-turbo spin echo, T1-weighted images ± contrast agent) of the lumbar spine demonstrated an intra- and extraforaminal space occupying lesion at the level L2/3 presumed to be a spinal neurinoma. Intraoperatively frozen section of the lesion revealed an epidural venous malformation. The definite histopathologic examination showed a cavernous malformation.

Conclusion: Cavernomas are rare spinal lesions, which may be difficult to diagnose preoperatively. When characteristic MRI features are missing, these lesions can be easily misdiagnosed as neurinomas. Complete surgical resection is warranted to prevent further neurological symptoms.

Keywords

spine; intra- and extraforaminal growth; vascular malformation; cavernoma

Abbreviations

CNS: Central nervous system; TSE: Turbo spin echo; SWI: Susceptibility weighted imaging

Introduction

The most common cause of lower back pain with or without radiculopathy are intervertebral disc protrusions or prolapses. Nevertheless, the differential diagnosis can be sometimes problematic including tumors like neurinomas, ependymomas or menigiomas, hematomas, epidural empyemas, synovial cysts or vascular malformations.

Capillary angiectasias, cavernous angiomas, arteriovenous malformations and venous angiomas are subtypes of the vascular malformations found in the central nervous system (CNS) [1]. Vascular malformations account for around 6-7% of all spinal tumors [2]. The incidence of cavernomas located in the CNS, including the brain and the spinal cord, is about 1.9 / 100,000 persons per year [3]. However,

taking into account only spinal cord cavernomas, these are even less common lesions with a reported prevalence of 5 - 12% of all intraspinal vascular malformations [4], with a prevalence of 3-5% of all spinal cord lesions [3, 5].

Cavernomas are vascular malformations composed of dilated sinusoidal venous channels lined with a single layer of endothelial cells without a complete vascular wall [6]. In majority, cavernomas arise from the vertebral bodiesand extend into the extradural space [7].

We describe a case of a lumbar extradural, intra- and extraforaminal cavernoma compromising the nerve root, causing lumbago and hypoesthesia. The lesion was preoperatively presumed to be a neurinoma. Intraoperatively, a frozen section revealed a cavernoma.

Case Presentation

Clinical findings: A 50-year-old woman sought medical attention due to progressive lumbago combined with a hypoesthesia of the upper part of the left leg. She described a subjective weakness of the left leg, which was operated twice in the past due to varices. No further neurological deficits were mentioned by the patient. The current neurological examination at the day of admission showed a hypoesthesia at the ventromedial area of the thigh of the left leg. No other neurological deficit, especially no paralysis, was observed.

Radiological findings: MRI of the lumbar spine showed a 11x23 mm sized extradural, intra- and extraforaminal lesion around the left nerve root L2 at the level of the second and third lumbar vertebral bodies with a "sand glass"shape. The lesion had an intermediate hyperintensity on T2-weighted images compared to the surrounding muscular tissue, almost isointensity on T1-weighted images, as well as a strong enhancement after administration of a contrast agent (Dotarem®, Guerbet, Sulzbach/Taunus, Germany). Shape and T2 signal intensity of the tumor were suitable for the diagnosis of a schwannoma, neurinoma or a meningioma. Those entities can show a strong contrast enhancement like the described formation (Figure 1).

Surgical treatment: A skin incision was performed over the left-sided paravertebral area at the level of the 2nd and 3rd lumbar vertebra followed by intramuscular preparation between the multifidus and longissimus muscles down to the spinal transverse processes L2 and L3. After that, intraoperative fluoroscopy confirmed the left sided isthmus of L2/L3. Removal of the intertransverse ligament revealed directly the lesion as well as the nerve root L2 and the spinal ganglion laterally from the lesion. The lesion was located dorsally of the nerve root extraforaminal, intraforaminal and in the lateral aspect of the spinal canal (Figure 2). The lesion showed a highly lobulated reddish appearance typical for cavernous angioma. Under microscopic assistance, delicate mircrodissection of the lesion from the nerve root and the dura was made using the bipolar coagulation forceps and micro dissectors. The spinal foramen was widened, removing partly the surrounding bone to increase access to the spinal canal. After complete removal of the cavernous malformation the intact dura was visualized as well as the exit of the spinal root nerve with no further signs of the lesion. After completed hemostasis closure of the wound was performed.

Histopathology: Histopathologic examination showed collagenous connective tissue with an angioma-like convolute consisting of dilated, thick- and thin-walled fibrotic blood vessels. An arterial differentiation could not be observed (Elastica-van-Gieson staining). Additionally, there were signs of

older hemorrhages indicated by the presence of iron containing macrophages (siderophages). These histological findings were consistent with a cavernous malformation. No signs of neurinoma-like tissue could be observed (Figure 3).

Postoperative course and Follow up: The patient recovered well without any new neurological deficits. The known hypoesthesia was described as less distinctive shortly after surgery. The MRI sequences with and without contrast enhancement 3 months after surgery showed a complete removal of the intra- and extraforaminal cavernoma.

Discussion

Spinal extradural cavernomas are rare lesions and difficult to diagnose preoperatively. Only a few cases have been published in the literature before [1, 8-15]. Intramedullary cavernomas are more common than the extra-/epidural localizations. Kivelev et al found 383 patients with intramedullary cavernomas in their review of the literature [10].

Cavernomas are defined as a subgroup of hemangiomas and are benign congenital malformations [16]. Although lacking mitotic activity, cavernomas grow throughout life, probably caused by recurrent hemorrhage and thrombosis with recanalization and organization of tissue [17].

Current classification schemes define them as vascular malformations, occurring throughout the body and CNS, which are clearly differentiated from true vascular neoplasms, such as haemangioblastomas.

The majority (about 80%) of all spinal epidural cavernous angiomas are located at the thoracic level dorsally from the spinal cord and may be present with myelopathy [18].

Lumbar epidural cavernous angiomas are preferably located in the ventral extradural space. Neurological symptoms caused by these cavernomas can mimic symptoms of other spinal pathologies including somatic pain or radiculopathy, sensory deficits, and weakness.

Sensory or functional deficits are most frequent (about 60%), followed by pain (34%) and bladder/bowel dysfunction (24 %) [19]. Ardeshiri et al were able to show in their retrospective study about spinal cavernomas that the postoperative Frankle Score of surgical treated patients was equal to the preoperative Frankle Score in 80% or improved in 20% of the cases [20].

Spinal venous malformations are mainly diagnosed by MRI [20]. MRI findings show heterogeneous signal intensities on T1- and T2-weighted images. Cavernomas are isointense on T1-weighted images and hyperintense on T2-weighted images presenting with homogenous contrast enhancement [15].

Hemosiderin deposits and degenerative signs of thrombosis and fibrosis often occur simultaneously. Extradural hemangiomas contain more often hemosiderin than intradural lesions, which may be presumably because of an easier degradation of blood products outside the blood–brain barrier [21]. Due to the small size of the involved vessels, arteriovenous shunting is often absent [21], hence digital subtraction angiography rarely shows AV-shunts in cavernomas.

Hemosiderin deposits, inhomogeneous T2-hyperintense and contrast enhancement mostly lead to the diagnosis of cavernomas based on MR images.

If characteristic MRI features of cavernomas are missing, these lesions can be misdiagnosed as disc herniation, meningioma, ependymomas, epidural hematoma, synovial cyst, or neurinoma similarly to the presented case.

The neurinoma-like appearance of the presented lesion on MR images with T2/STIR hyperintensity was misleading. However, due to the high signal intensity in the T2-weighted images and the strong contrast enhancement, a vascular malformation could have been considered as a differential diagnosis. Additional MRI sequences like the susceptibility weighted imaging (SWI) and the conventional gradient echo,T2*-weighted MRI sequence, could be made to verify the differential diagnosis of a vascular malformation showing hemosiderin deposits in the area of interest.

Based on the misleading MRI appearance and well known results of the postoperative outcome, comparable lesions should be extirpated to receive a histologically proved diagnosis and to prevent worsening of neurological symptoms.

Conclusion

Spinal cavernomas are rare lesions and sometimes difficult to diagnose radiologically. These lesions can be easily misdiagnosed as a neurinoma, ependymoma, meningioma, hematoma, or a synovial cyst in MRI scans. Surgical resection is the suitable therapy of spinal cavernomas to confirm the right diagnosis and to prevent further neurological symptoms and damage.

Figures

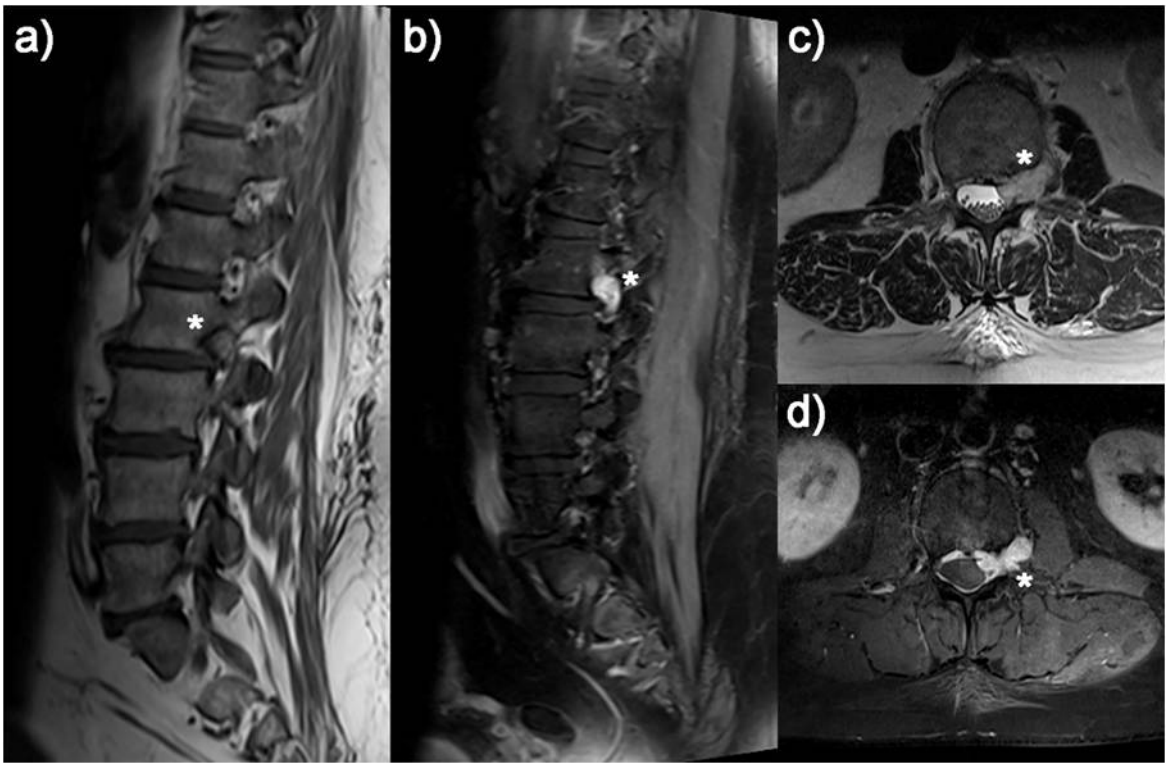

Figure 1: T1-turbo spin echo (TSE), sagittal plane a) showing a nearly isointense intraforaminal lesion at the left neuroforamina of the 2nd and 3rd lumbar vertebra. Transversal T2-weighted TSE c) showing a hyperintense extradural, intra- and extraforaminal lesion around the left nerve root L2. T1-weighted TSE images with contrast enhancement in transversal b) and sagittal d) planes show the hourglass shaped tumor with strong, almost homogenous enhancement.

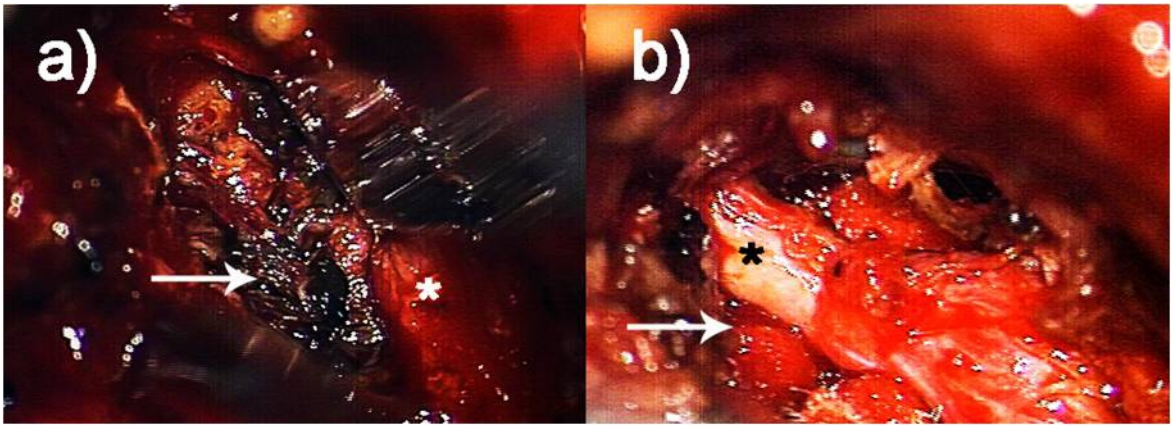

Figure 2: The lesion (→) was located dorsally of the nerve root (*) extraforaminal, intraforaminal before a) and after resection b).

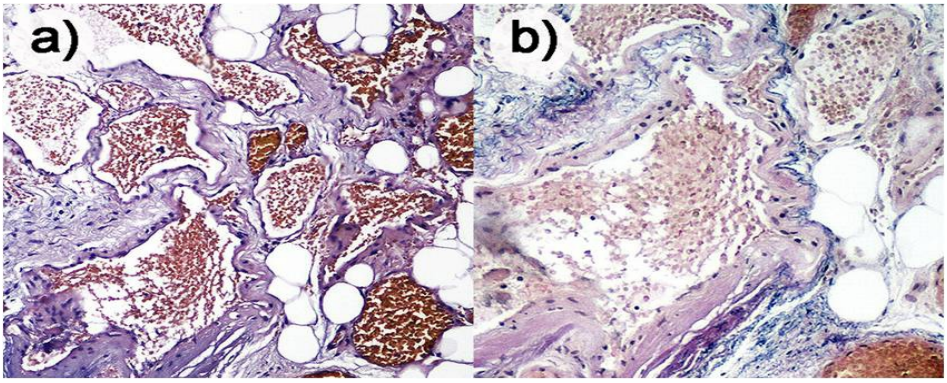

Figure 3: Complex of dilated, thin- and thick- walled fibrotic blood vessels a) Hematoxylin-eosin staining without arterial differentiation in terms of elastic laminae b) Elastica-van-Gieson staining with a 100 fold magnification.

Author's Contributions

Oliver Gembruch was a major contributor in writing the manuscript and was part of the neurosurgical team. Haemi Phaedra Schemuth analyzed the images and was also a contributor in writing the manuscript. Sarah Teuber-Hanselmann performed the pathological examination and was also a contributor in writing the manuscript. Yayha Ahmadipour performed the neurological examinations. Elias Lemonas performed the neurosurgical procedure and was a contributor in writing the manuscript. All authors read and approved the final manuscript.

References

1. Lee JW, Cho EY, Hong SH, Chung HW, Kim JH, Chang KH, et al. Spinal epidural hemangiomas: various types of MR imaging features with histopathologic correlation. AJNR Am J Neuroradiol. 2007; 28: 1242–1248.

2. Ruíz DS, Yilmaz H, Gailloud P. Cerebral developmental venous anomalies: current concepts. Ann Neurol. 2009; 66: 271–283.

3. El-Koussy M, Stepper F, Spreng A, Lukes A, Gralla J, Brekenfeld C, et al. Incidence, clinical presentation and imaging findings of cavernous malformations of the CNS. A twenty-year experience. Swiss Med Wkly. 2011; 13; 141: w13172.

4. Cosgrove GR, Bertrand G, Fontaine S, Robitaille Y, Melanson D. Cavernous angiomas of the spinal cord. J Neurosurg. 1988; 68: 31-36.

5. Padovani R, Tognetti F, Proietti D, Pozzati E, Servadei F. Extrathecal cavernous hemangioma. Surg Neurol. 1982; 18: 463–465.

6. Clatterbuck RE, Eberhart CG, Crain BJ, Rigamonti D. Ultrastructural and immunocytochemical evidence that an incompetent blood-brain barrier is related to the pathophysiology of cavernous malformations. J Neurol Neurosurg Psychiatry. 2001; 71: 188-192.

7. Rovira A, Rovira A, Capellades J, Zauner M, Bella R, Rovira M. Lumbar extradural hemangiomas: report of three cases. AJNR Am J Neuroradiol. 1999; 20: 27-31.

8. Petridis AK, Doukas A, Hugo HH, Barth H, Mehdorn HM. A rare case of extradural lumbar nerve root cavernoma. Eur Spine J. 2011; 20 Suppl 2: 348-349.

9. Akhaddar A, Oukabli M, En-Nouali H, Boucetta M. Acute postpartum paraplegia caused by spinal extradural capillary hemangioma. Int J Gynaecol Obstet. 2010; 108: 75–76.

10. Kivelev J, Niemelä M, Hernesniemi J. Outcome after microsurgery in 14 patients with spinal cavernomas and review of the literature. J Neurosurg Spine. 2010; 13: 524-534.

11. Vassal F, Péoc'h M, Nuti C. Epidural capillary hemangioma of the thoracic spine with proximal nerve root involvement and extraforaminal extension. Acta Neurochir. 2011; 153: 2279–2281.

12. Seferi A, Alimehmeti R, Vyshka G, Bushati T, Petrela M. Case study of a spinal epidural capillary hemangioma: a 4-year postoperative follow-up. Global Spine. 2014; 4: 55–58.

13. Fontaine S, Melanson D, Cosgrove R, Bertrand G. Cavernous hemangiomas of the spinal cord: MR imaging. Radiology. 1988; 166: 839–841.

14. Morioka T, Nakagaki H, Matsushima T, Hasuo K. Dumbbell-shaped spinal epidural cavernous angioma. Surg Neurol. 1986; 25: 142–144.

15. Gupta S, Kumar S, Banerji D, Pandey R, Gujral R. Magnetic resonance imaging features of an epidural spinal hemangioma. Australasian Radiol. 1996; 40: 342–344.

16. Hurt M. Les hemangioblastomes intrarachidiennes. Neurochirurgie. 1975; 21: 1–136.

17. Gutkelch AN. Hemangiomas involving the spinal epidural space. J Neurol Neurosurg Psychiatry. 1948; 11: 199–210.

18. Aoyagi N, Kojima K, Kasai H. Review of spinal epidural cavernous hemangioma. Neurol Med Chir. 2003; 43: 471–475.

19. Badhiwala JH, Farrokhyar F, Alhazzani W, Yarascavitch B, Aref M, Algirid A et al. Surgical outcomes and natural history of intramedullary spinal cord cavernous malformations: a single-center series and meta-analysis of individual patient data. J NeurosurgSpine. 2014; 21: 662–676.

20. Ardeshiri A, Özkan N, Chen B, Stein KP, Miller D, Hütter BO, et al. A retrospective and consecutive analysis of the epidemiology and management of spinal cavernomas over the last 20 years in a single center. Neurosurg Rev. 2016;39: 269-276.

21. Leitlinien für Diagnostik und Therapie in der Neurologie; 4. überarbeitete Auflage 2008, S. 654 ff, ISBN 978-3-13-132414-6; Georg Thieme Verlag Stuttgart.

22. Graziani N, Bouillot P, Figarella-Branger D, Peragut JC, Dufour H, Grisoli F. Cavernous angiomas and arteriovenous malformations of the spinal epidural space: report of 11 cases. Neurosurgery. 1994; 35:856–864.

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