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Thoracic Intraspinal Venous Angioma Mimicking a Schwannoma: A Case Report

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Abstract

Introduction: A spinal epidural venous angioma is a very rare entity and can be misdiagnosed easily. The epidural venous angioma can cause neurological symptoms due to the affection of the nerve root and should be removed before irreversible deficits occur.

Case Presentation: We present an uncommon case of a 42-year-old woman with a left sided intraspinal lesion in the thoracic level of T2/3, which was presumptively diagnosed as a schwannoma in the MRI. Intraoperatively, the lesion turned out to be an epidural venous malformation which affected the nerve root. The histopathologic examination showed a venous angioma.

Conclusion: Epidural venous malformations are rare lesions and difficult to diagnose preoperatively. When missing typical MRI features, these lesions can be easily misdiagnosed as schwannomas. Surgical resection is warranted to prevent further neurological symptoms or myelopathy.

Keywords

Venous angioma; Venous malformation; Tumor; Spine; Extradural growth

Introduction

Nervous symptoms like back pain with unilateral or bilateral radicular pain or a hypaesthesia are mainly caused by degenerative spine disease. Nonetheless, differential diagnoses includes tumor, hematoma, abscess, synovial cyst and vascular malformation.

Vascular malformations are common lesions in the central nervous system and can be divided into capillary angiectasias, cavernous angiomas, arteriovenous malformations and venous angiomas [1]. Venous angiomas are the most frequently encountered cerebral vascular malformation with an incidence of up to 2.6% in a series of 4069 brain autopsies [2] and account for around 15% of the vascular malformations [2, 3]. However, vascular malformations account for around 6-7% of the spinal tumors [4, 5]. Spinal, extradural vascular malformations are rare and completely extradural venous angiomas are even more uncommon [1, 6]. The definitive diagnosis is usually made by surgery.

We describe a case of a thoracic epidural mass which was preoperatively diagnosed as a schwannoma in the magnetic resonance imaging (MRI) but intraoperatively identified as an epidural venous angioma.

Case Presentation

Clinical Findings: A 42-year-old woman sought medical attention with a left sided hemihypaesthesia which occurred two years earlier without any spinal trauma in the past. There was no other disease except of a borreliosis infection suffered 7 years ago and cured without any following symptoms. Further neurological deficits were not mentioned by the patient. In the past several neurological examinations were performed by different neurologist without any findings. Lumbar puncture revealed no signs for any infection in the cerebral fluid. Electrophysiological examinations were also normal. The current neurological examination at the day of the admission showed no focal or general neurological deficit.

Radiological Findings: MRI of the spinal axis was performed at an outside clinic for further diagnostic work up. MR revealed a sharply demarcated "sand glass" shaped lesion in the neuroforamen at the level of the second and third thoracic vertebra with a hyperintense signal on T2 and STIR. The lesion was located in the left lateral epidural space with slight spread cranially and caudally and slight impression of the dural sac and comprising the spinal cord without any signs of myelopathy, and a beginning extraforaminal growth. The lesion showed a homogenous hypointense T1 signal with an intense homogenous contrast enhancement [Fig. 1]. The neuroradiological diagnosis was a schwannoma with compression of the dural sac.

Surgery: The patient underwent surgical resection of the lesion with a left-sided hemilaminectomy of the vertebra T3 under electrophysiological monitoring. A further cranial and caudal osseous decompression followed. After decompression the epidural lesion was seen. Intraoperatively, the lesion presented as an epidural angioma located on the dura, embedding the 3rd thoracic nerve root with mass effect on surrounding structures. A haemostasis procedure and subsequent removal of the lesion followed, using bipolar coagulation. The dura was opened, showing that the lesion was restricted to the extradural space.

Histological Findings: Histopathologic examination showed dural tissue with an adjacent angioma-like convolute consisting of thick- and thin-walled fibrotic blood vessels lined by a single layer of CD31-positive endothelial cells. No distinct arterial differentiation was observed (Elastica-van-Gieson staining). These histological findings revealed a venous malformation.

Outcome: The postoperative MRI showed complete removal of the venous malformation. The patient recovered well without any new neurological deficits. The known hemihypaesthesia was regredient as well. The MRI sequences with and without intravenous contrast application after 9 months showed a complete removal of the epidural venous malformation [Fig. 2]. The patient did not mention any residual symptoms.

Discussion

Epidural venous malformations such as venous angiomas are rare lesions and difficult to diagnose preoperatively. Only a few cases are published in the literature before [1, 6-8]. Decker et al. presented the first case of a spinal lumbar venous angioma in 1978 [6]. Five further cases of spinal venous angioma are published over the last years [1, 7, 8], but only one thoracic venous angioma was described before [7].

Venous angiomas are described as a cluster of venous radicles that converge into a collecting vein [1]. As in the present case, the H&E staining showed the irregular and cystic vessels with thin walls and surface-lining epithelial cells, typically for the venous angioma. Furthermore, immunohistochemical analysis revealed that the surface-lining epithelial cells were positive for CD31.

Neurological symptoms caused by the venous angiomas are similar to those of the other spinal pathologies. The clinical presentation includes somatic pain or radiculopathy, sensory deficits and weakness. Five of every six affected patients recorded somatic pain or radiculopathy [1, 6-8], and one of them showed weakness of the left upper limb [8].

In most cases MRI can reliably diagnose venous malformations. The MRI signals in T1-weighted and T2-weighted images of venous angiomas range from isointense to hyperintense. Venous angiomas can show no or irregularly heterogeneous enhancement with well-defined tumor margins. There are no flow voids seen on MRI [9].

When missing these typical MRI features, these lesions can be misdiagnosed as herniated disc prolapse, tumor, hematoma or synovial cyst [1, 6-8, 10].

In the presented case the lesion showed a typical schwannoma-like appearance with a homogenous strong contrast enhancement, a sharp delineation and a hyperintensity in T2/STIR and was misinterpreted as a schwannoma due to the striking similar appearance in MRI.

As a result of the difficulty finding of the correct diagnosis, the lesion should be extirpated to receive a histologically proved diagnosis and to prevent further worsening of the tumour entity or a further neurological symptoms.

Conclusion

Spinal, epidural venous angiomas are rare lesions and difficult to diagnose preoperatively. In MRI scans these lesions can be easily misdiagnosed as tumor, hematoma or a synovial cyst. Surgical resection is warranted to receive the correct diagnosis and to prevent further neurological symptoms or myelopathy.

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Author(s) Contributions

Oliver Gembruch was a major contributor in writing the manuscript and was part of the neurosurgical team. Cornelius Deuschl 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. Tobias Schoemberg was part of the neurosurgical team. Neriman Özkan performed the neurosurgical procedure. All authors read and approved the final manuscript.

Figures

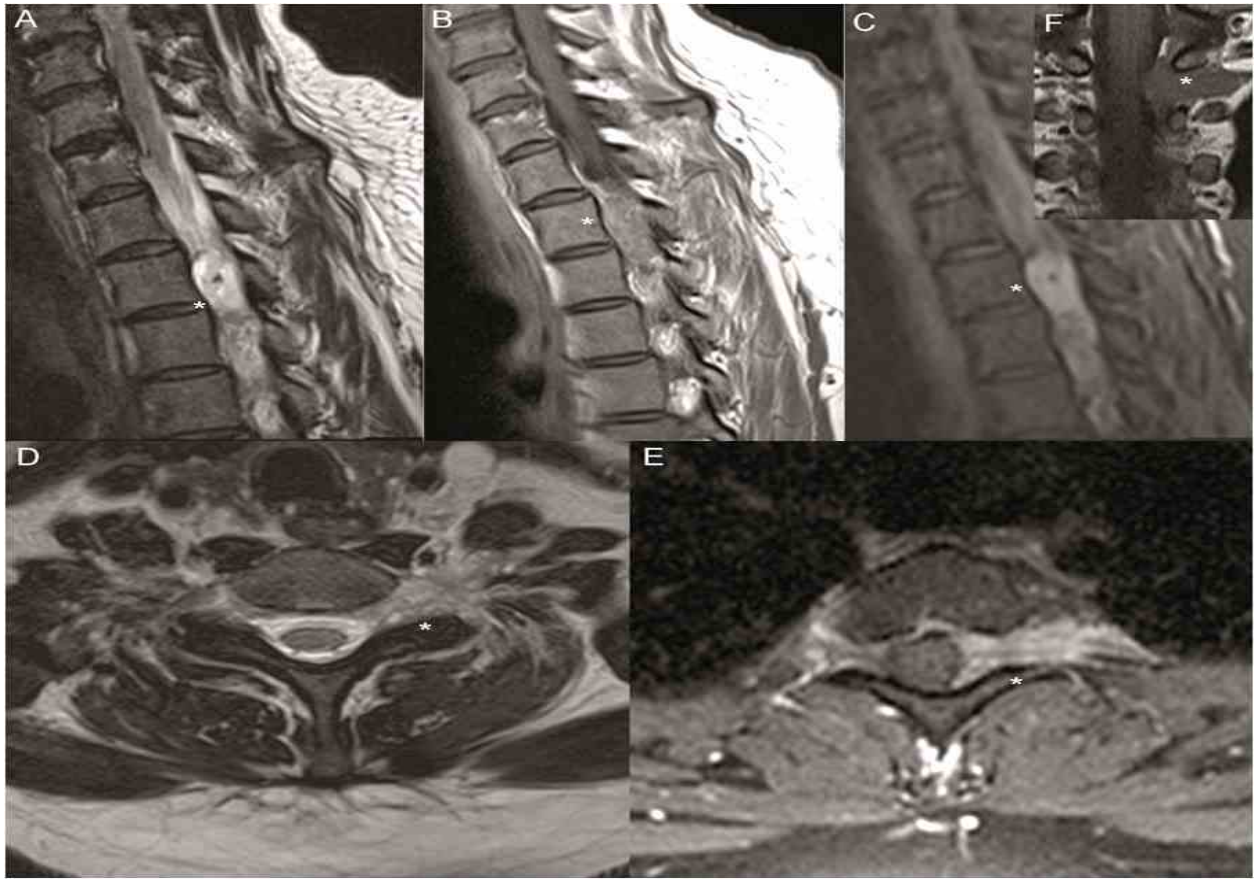

Figure 1: T2-weighted (A, D) and STIR (C) images show a hyperintense lesion with a dull hyperintensity of the bordering myelon (D). The mass is clearly delineated in T1 (B, E, F) and T2. It shows a strong homogenous contrast enhancement (B, E). Dilated vessels or flow-voids were not definable.

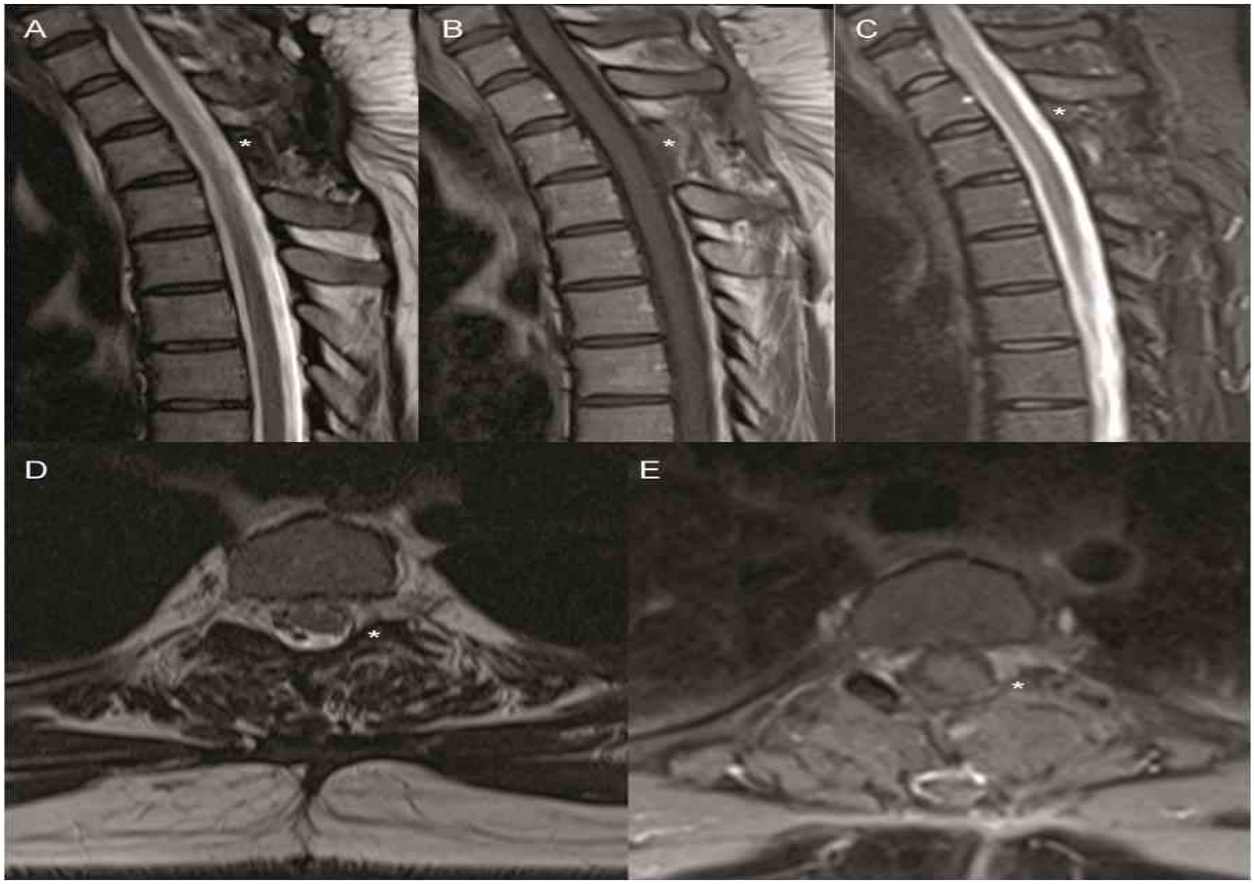

Figure 2: MRI-Scan 9 months after resection. T2-weighted images (A, D), T1-weighted images (B, E) and the STIR (C) do not show any signs of tumor recurrence.

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