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Thoracic spinal angiomyolipoma: case report from an Ethiopian neurosurgical teaching hospital. Illustrative case

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BACKGROUND Angiomyolipoma (AML) of the spine is a rare benign neoplasm (accounting for 0.14%–1.2% of all spine tumors) that is often described along with angiolipoma because of their similarities. They occur almost exclusively in the extradural space, with the thoracic spine being the commonest level.

OBSERVATIONS The authors present the clinical presentation, diagnosis, and treatment of an extremely rare case of thoracic spine AML in a 47-year-old male patient. The patient underwent laminectomy and gross total resection of the tumor and had an excellent immediate postoperative neurological recovery and long-term functional neurological outcome.

LESSONS It is always wise to consider rare benign spinal epidural neoplasms such as spinal AMLs in the differential diagnosis of spinal epidural mass, despite metastasis being the commonest epidural tumor with variable modes of treatment, because the management of benign spinal epidural masses such as spine AML is always surgical and associated with an excellent long-term outcome.

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KEYWORDS angiolipoma; angiomyolipoma; case report; spinal epidural mass; spinal tumors

Angiomyolipoma (AML) of the spine is a rare benign neoplasm causing a wide array of neurologic deficits. AMLs usually occur in the kidneys. They are usually grouped together with angiolipoma (AL) due to its histopathologic similarities. AMLs differ from angiolipomas by their abundant smooth muscle component, which is absent in angiolipomas.[1–5] They account for 0.14%–1.2% of all spine tumors.[1] This illustrative case describes an extremely rare case of thoracic angiomyolipoma, including the patient's presentation, diagnosis, treatment, and outcome.

Illustrative Case

A 47-year-old male patient presented to our hospital with a complaint of progressive lower extremity weakness of 1 year's duration. Otherwise, he had no incontinence.

His physical examination revealed hypertonic lower extremities with hyperreflexia. The muscle strength in the lower extremities was 4/5 on the right side and 2/5 on the left side. He had hypoesthesia below the T6 dermatome level but no other significant sensory deficit.

Imaging Findings

Magnetic resonance imaging (MRI) revealed a T2 and T1 hyperintense elliptical mass measuring 2.8 × 1.3 × 1.6 cm on the dorsal aspect of the spinal canal at the T6–7 level. Very small central hypointensities were observed on the T2 sequence. The mass had an extension to the left neural foramen and was displacing the cord to the right. It also showed postcontrast enhancement (Fig. 1).

Surgery

A T6–8 laminectomy was performed with fluoroscopic guidance with the patient under general anesthesia and in prone position. An elongated extradural mass with a whitish pearly part extending to the neural foramen of the T6 spine was found. The mass occupied the entire extradural space and pushed the theca to the right side.

ABBREVIATIONS AL = angiolipoma; AML = angiomyolipoma; MRI = magnetic resonance imaging.

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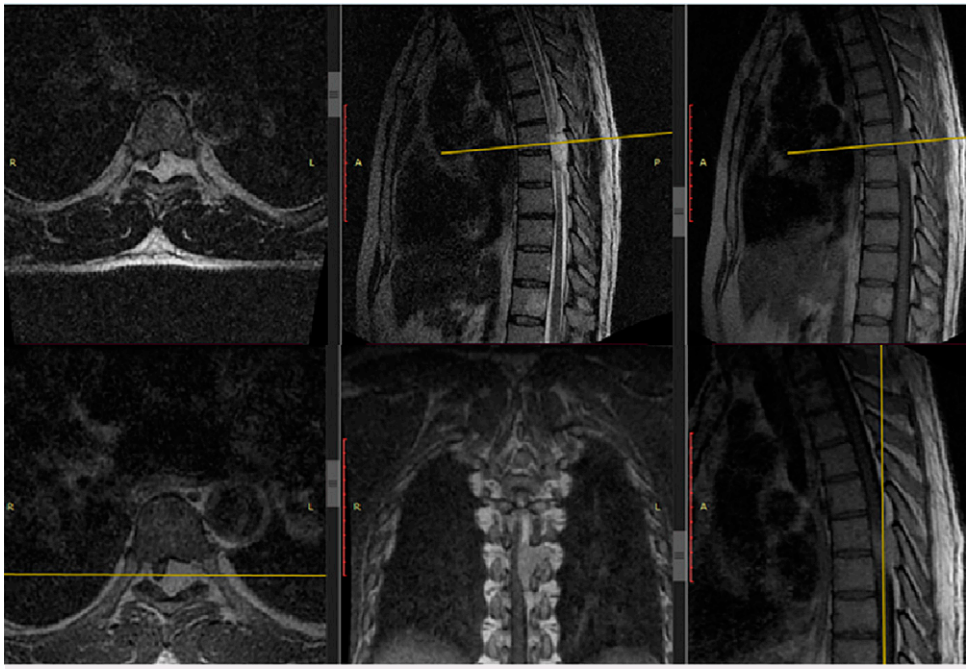

FIG. 1. MRI sequence of a midthoracic (T6–7) epidural mass that is hyperintense to both T1 and T2 with postcontrast enhancement.

The tumor surface was cauterized and meticulously separated from the dorsal dural surface. It was followed to the neural foramen, and gross total resection of the tumor was achieved.

Postoperative Course

The patient started physiotherapy on the second postoperative day. He was able to walk by himself without any support in his second postoperative week, and he currently is on his fifth smooth postoperative year after the surgery with intact long-term neurological function.

Histopathologic Result

The histopathologic result showed a soft tissue mass composed of mature adipocytes, blood vessels, mesenchymal tissue, and smooth muscle cells (see Fig. 2 for gross pathology and Fig. 3 for histopathology).

Discussion

Observations

Spine angiomyolipoma is rare with an incidence of 0.14%–1.2%.[1] Overall, it most commonly involves the thoracic spine (78%), the lumbar column (10%), and the cervical or sacral column (<1%).[6] The exact pathogenesis of AMLs is unknown. However, there are two major explanations mentioned for the origin of these rare benign tumors: pluripotent mesenchyme cells giving different mesenchyme cells versus congenital malformation classifying them as true hamartomas.[7–9]

Patients with AMLs or AIs most frequently present with long-standing back pain, which then develops into progressive neurological symptoms.[8,9] The mean duration of symptom progression at diagnosis is 1 year.[10] Fast progression (acute) neurological symptoms might occur in some of the cases of these tumors and is attributed to vascular factors such as venous stasis with thrombosis, intratumoral bleeding, or steal phenomenon leading to an acute cord compression.[6,11–14]

On MRI, the fatty content appears hyperintense on both T1- and T2-weighted images and hypointense on fat-suppressed images, whereas the vascular component appears hypointense on T1-weighted and hyperintense on T2-weighted images and shows intense enhancement in postcontrast images.[2,15,16] However, infiltrative properties such as invasion to adjacent bone or extension to the perilesional space and the masses prone to the ventral location have been mentioned as distinctive features of AML compared with major patterns of angiolipoma.[4]

Differential diagnosis of AML includes fat-containing tumor (such as lipoma, which presents with a typical Y configuration with circumferentially

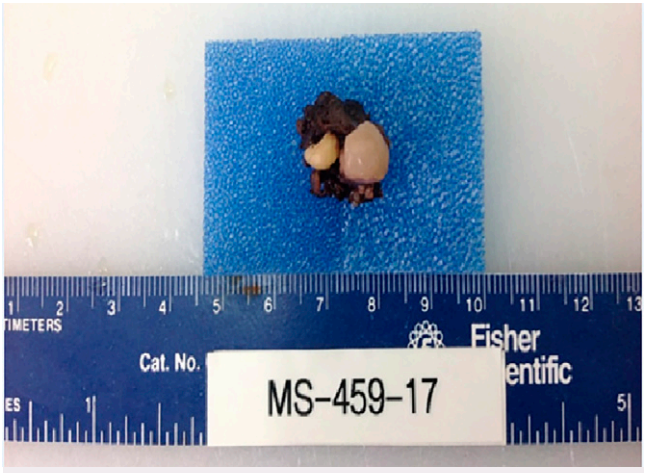

FIG. 2. Gross tissue appearance of angiomyolipoma: A light-colored lipoma and darker smooth muscle and vascular component are seen.

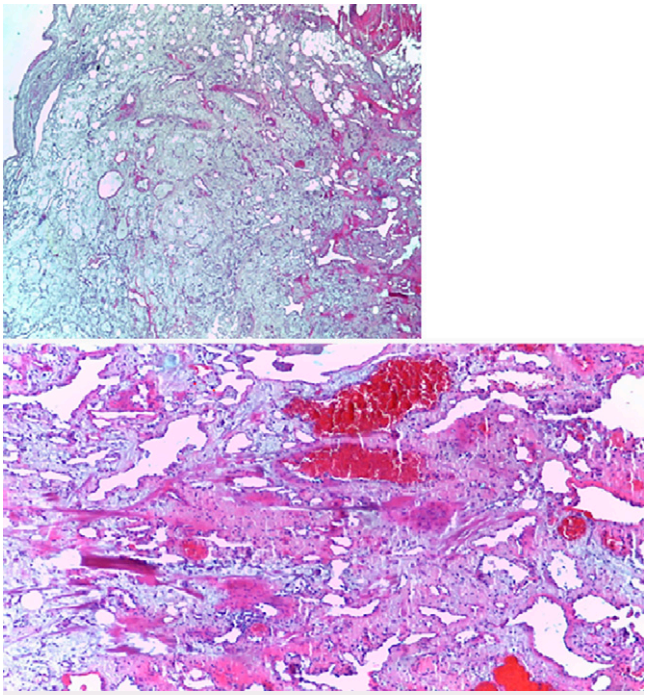

FIG. 3. Histology of angiomyolipoma (**upper** is under low magnification [×10], and **lower** is under high magnification [×40] with hematoxylin and eosin stain) composed of mature fat cells, smooth muscle cells, and abnormal vascular elements.

compressed dural sac and shows no definite contrast enhancement pattern, and liposarcoma, which frequently has irregular thick-ended septa), prominently vascular tumor (such as spinal epidural hemangioma, which has T2-low signal intensity rim due to fibrous capsule or hemosiderin deposition, multisegment involvement, and rare T1-high signal intensity foci [except combined internal hemorrhage]), and other T1-high signal intensity lesion (such as subacute stage epidural hematoma, which lacks fat suppression sequence that can be used to distinguish hemorrhage from the true fat component).[17–19]

The mainstay of management for AMLs is gross total resection of the tumor for noninfiltrating tumors and subtotal excision for infiltrating ones, respectively, with an excellent outcome in both cases.[9,15] No adjuvant treatment is indicated.[6]

Lessons

Although metastasis is the commonest spinal epidural tumor with variable management and outcome, it is wise to consider rare benign spinal epidural neoplasms in the differential diagnosis because the management is always surgical and associated with an excellent outcome.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: all authors. Acquisition of data: Laeke, Yibeltal. Analysis and interpretation of data: Laeke. Drafting the article: Laeke, Yibeltal. Critically revising the article: Laeke, Yibeltal. Reviewed submitted version of manuscript: Laeke, Yibeltal. Approved the final version of the manuscript on behalf of all authors: Laeke. Administrative/technical/material support: Laeke. Study supervision: Kwon.

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