Spinal Angiolipoma

—Case Report—

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Abstract

A 69-year-old male presented with a rare spinal angiolipoma manifesting as history of back pain, and numbness in both lower limbs, which progressed over a period of 5 years. Total T10–T12 laminectomy was performed and the tumor was removed en bloc. The symptoms gradually improved postoperatively. Spinal angiolipoma is an uncommon benign extradural tumor of spine, which accounts for 0.14–1.2% of all spinal tumors and is a rare cause of spinal cord compression. Recognition of this entity is crucial as a benign and curable cause of paraplegia and back pain.

Key words: angiolipoma, lipoma, thoracic spine, tumor

Introduction

Angiolipoma of the spine is a benign neoplasm consisting of both mature fatty tissue and abnormal vascular elements. The first report of spinal angiolipoma was found in a 16-year-old boy with an unsuccessfully treated spinal thoracic mass which was confirmed at autopsy in 1890.⁷⁾ Since then angiolipoma has been considered to be a vascular variant of lipoma.³⁾ Angiolipoma was described as a distinct anatomopathologic entity in 1960.¹⁴⁾ The estimated occurrence is 0.14–1.2% of all spinal tumors, and only 2–3% of extradural spinal tumors.³⁾ Sometimes the tumor may be more aggressive and invade the vertebral body and adjacent soft tissues.¹⁷⁾ We report another case of spinal angioma in an elderly patient.

Case Report

A 69-year-old male presented to our outpatient clinic with a 5-year history of back pain, bilateral progressive worsening paresthesias, and hypesthesia below the level of umbilicus. The paresthesias spread slowly upward, beginning from the foot. He denied any motor weakness and bowel or bladder symptoms. Neurological examination revealed decreased sensation to pin prick and light touch below the T9–T10 intervertebral level as well as poor joint position sense in the lower limb. No atrophy or fasciculation was noted, and examination of the motor system revealed no abnormality. Spinal examination was unremarkable with no local tenderness or deformity.

Computed tomography (CT) showed a dorsal hypodense mass extending from the T9 to T11 levels. Magnetic

resonance (MR) imaging of the thoracic spine revealed a 7-cm elongated mass in the posterior epidural space extending from the T9 to T11 levels. Prominent cord compression was noted at the T10–T11 levels. The mass appeared as slightly high intensity with areas of hypointensity at the T10 level on T₁-weighted images (Fig. 1A). T₁-weighted imaging with gadolinium showed inhomogene-

Fig. 1 A: Preoperative sagittal T₁-weighted magnetic resonance (MR) image showing a T9–T11 extradural mass as slightly high intensity with areas of hypointensity at the T9 level, causing maximum compression at the T10 level. B: Preoperative sagittal T₁-weighted MR image with contrast medium showing inhomogeneous enhancement. C: Preoperative axial T₁-weighted MR image at the T10 level showing a hyperintense extradural mass. D: Postoperative axial T₁-weighted MR image at the T10 level confirming total resection of the tumor.

Fig. 2 Photomicrograph of the tumor tissue showing mature adipose tissue interspersed with blood channels of various sizes, indicative of angioliipoma. Hematoxylin-eosin stain, original magnification $\times 100$.

ous enhancement at the T10 level (Fig. 1B, C). The differential diagnosis included lipomatosis, metastasis, chronic extradural hematoma, lymphoma, angioliipoma, nerve sheath tumor, and meningioma.

Total T10–T12 laminectomy using high speed drill revealed a dark yellow, globular firm mass with minimal vessels on its surface. The mass appeared different to the regular epidural fat tissue and was easily dissected from the dura. The dura was not opened and the mass was removed en bloc. No residual tumor was identified caudal and rostral to the removed fatty tumor. Postoperative MR imaging confirmed the total removal of tumor (Fig. 1D). Postoperative course was uneventful with marked neurological improvement. Gradual improvement in hypesthesia was noted during the follow up.

Histological examination of the removed specimen was consistent with angioliipoma. The characteristic features of mature adipose tissue without pleomorphic or myxoid degeneration were found, interlaced with few vascular elements ranging from distended capillaries, venules, and blood-filled lakes to small arterioles (Fig. 2). Immunostaining found Ki-67 labeling index of $< 1\%$, no atypia of the endothelial cells, and no mitotic figures, consistent with benign lesion.

Discussion

An exhaustive review of the world literature identified 121 cases of extradural spinal angioliipoma from 1890 to 2009. Of the 121 reported patients, 72 were females and 49 were male. The mean age of occurrence was 44.03 ± 5.9 years, but 17 of the 121 cases including our case occurred in the elderly population.¹²⁾

Angioliipomas manifest as signs and symptoms consistent with other benign space-occupying lesions of the spine. The most common initial symptom is progressive or sudden weakness in 51 patients (42.1%), thoracic or lumbar pain in 28 (23.1%), and alteration in sensation in 24 (19.8%). However, uncommon presentations included gait disturbances in 2 patients^{15,30)} (1.7%), unilateral or bilateral sciatica in 6 (5%), congenital occurrence in 1, and no

symptoms in 9.^{4,12)} The symptoms usually evolve over a period of months to years. The mean interval between initial symptom and diagnosis was 20.2 months. Nineteen of the 121 patients had duration of symptoms more than 36 months.¹²⁾ Indeed, in our case, the duration of symptoms was 60 months, exceeded by the 27-year duration of symptoms in previous reports.²³⁾ A relapsing and remitting course mimicking demyelinating disorder was reported in 8 of 121 (6.6%) cases.^{4,10,29)} Regardless of the duration, rapid deterioration has occurred in a few cases due to factors such as venous thrombosis, tumoral hemorrhage, vascular steal phenomenon, and intratumoral abscess. Spinal epidural angioliipoma is rarely complicated by intratumoral abscess,²⁴⁾ posttraumatic bleeding and venous thrombosis, and vascular steal phenomenon^{1,18)}; leading to rapid deterioration and subacute paraparesis.

Histologically, angioliipoma is considered to be a hamartoma consisting of mature fat cells with vascular proliferation, described as being either normal or mimicking capillary angioma, cavernous angioma, or arteriovenous malformations.²¹⁾ Normal karyotypes in a study of 20 patients with angioliipoma suggest the lesion is reactive or hamartomatous.²⁷⁾ Angioliipoma may constitute an intermediate entity in the spectrum of hemangiomas and lipomas. The more invasive infiltrating type of spinal angioliipoma would then represent a shift towards the hemangioma end of the spectrum. Tumors with an abundance of smooth muscle proliferation are further subclassified as angiomyolipomas.¹⁰⁾ Angioliipoma can be classified into noninfiltrating and infiltrating types.²¹⁾ The former type is more common and is well encapsulated. Histologically, the infiltrating angioliipoma consists of mature lipocytes with delicate, proliferating blood vessels wedging haphazardly into the surrounding tissue.¹⁷⁾ The ratio of fat to vessels is variable and ranges from 1:3 to 2:3. Atypia, pleomorphism, mitotic figures, or karyotypic abnormality were never found.¹²⁾ Despite the infiltrating and often worrisome proliferative nature of infiltrative angioliipoma, malignant transformation and neural tissue infiltration have never been reported. Additionally, low proliferation rates are found by Ki-67 immunostaining which favors a benign course.¹⁸⁾

The histogenesis of angioliipoma is poorly understood, but several theories have been proposed. Angioliipoma may originate from the primitive pluripotent mesenchymal cells, and early inclusion of pluripotent stem cells during the developmental ossification of neural arch is believed to be a prerequisite for spinal angioliipoma.⁹⁾ The strong predilection of infiltrating angioliipoma for anterior localization is thought to be due to the early inclusion of the immature pluripotent stem cells in the ventral and lateral spinal canal.¹⁰⁾ The concept of interperiosteodural space in the development of angioliipoma was recently introduced.¹¹⁾ This space represents an anatomical continuum extending from the coccyx to the orbit, and contains fat tissue that is abundant at the level of the orbit and the epidural spinal space, and sparser at the level of the cavernous spaces. Therefore, angioliipomas may represent a characteristic tumor that illustrates the interperiosteodural concept because angioliipomas essentially develop in

the fat tissue contained in these spaces. This dynamic concept might explain the occurrence of this tumor along the craniospinal axis. Predisposing factors are not clearly established, although a possible hormonal influence on the development, maintenance, or progression of angiolipomas is suggested by predominance in females, and aggravation of symptoms with acute presentation during pregnancy. Obesity is occasionally reported in patients with spinal angiolipomas.^{4,12)}

The most common location of spinal angiolipoma is the thoracic epidural space. Thoracic predominance of the tumor could be the result of the regional variation in blood supply in the spine, in particular that the midthoracic spine is least perfused.¹⁸⁾ Nevertheless, angiolipoma has been reported as cervical in 1.6%, lumbar in 9.6%, sacral in 0.8%, cervicothoracic in 4.1%, thoracolumbar in 2.4%, lumbosacral in 2.4%, and intracranial in 12 cases.^{4,11,17,19)} Infiltrating type of angiolipoma occurred in 21 cases (17.4%) of spinal angiolipoma.^{4,12)} Infiltration of the vertebral body was more often associated with anterior or anterolateral localization (12.4%) than posterior localization (5%).

CT demonstrates angiolipoma as hypodense and provides information on the degree of bony involvement. CT may not be specific for spinal epidural angiolipomas and could be misleading.²³⁾ MR imaging is considered the gold standard imaging modality.^{2,12,25)} MR imaging appearances of noninfiltrating spinal angiolipomas are hyperintensity and often inhomogeneous intensity on T₁-weighted images. The degree of central hypointensity on T₁-weighted images is predictive of the degree of vascularity. Significant heterogeneity in the imaging studies is attributed to the variable vascular and adipose elements of the tumor.²⁵⁾ In our case, the T₁-weighted images were less hypointense, consistent with few vascular elements. On T₂-weighted images, the tumors have variable intensity, with hyperintensity being more common. Gadolinium enhancement is due to the vascularity of these tumors. This phenomenon allows differential diagnosis between extradural lipomatosis and spinal angiolipoma, as the former is not enhanced.²²⁾ Gadolinium enhancement might be undetected due to the hyperintensity of angiolipomas on T₁-weighted imaging, but gadolinium infusion with fat suppression sequences may enhance areas of abnormal signal intensity within the fatty tumors.²⁰⁾

Spinal angiolipomas are treated exclusively by surgical removal of the lesion. Most extradural non-infiltrative tumors are amenable to complete excision via laminectomy. Spinal angiolipomas appear to be well-circumscribed lesions with no adhesions to the dura. A total of 112 patients underwent surgery and gross total resection was carried out in 83 cases (74.1%), subtotal resection in 6 (5.4%), biopsy sampling in 1 (0.8%), no mention of the extent of removal in 22 (19.6%), and no mention of surgery in 5 cases.^{4,10,12)} Preoperative embolization had been performed prior to T9 corporectomy and resection.²⁶⁾ Finally, radiotherapy was undertaken in only three cases.^{5,8,13)} Total removal of infiltrating angiolipomas, that often involve the vertebral body, has been recommended using the anterolateral approach and stabilization of the affected

vertebrae is desirable.²⁹⁾ Extent of tumor resection in infiltrative angiolipoma has been debated, but most authors agree that risking neurological function is not necessary with aggressive attempts to attain gross total removal.^{5,10,12,29)} Further, the invaded vertebral body could be preserved, as analogous to vertebral hemangiomas, spinal angiolipomas may not enlarge. Postoperative radiotherapy is not generally favored.⁵⁾

The postoperative course ranged from 3 months to 10 years. Good outcomes were obtained after subtotal resection in 8 of 11 cases of infiltrating angiolipoma.²⁸⁾ Tumor recurrence following surgery is rare, and only two cases (1.8%) were reported: one after gross-total removal⁶⁾ and one infiltrating tumor following subtotal resection.¹⁷⁾ The earlier reports record a total of six deaths, with no clear indication of cause of death^{4,10,12)}; but two deaths (1.8%) were reported recently in patients with non-infiltrating angiolipomas. The causes of mortality were cervical glioblastoma¹⁶⁾ and respiratory infection.¹⁴⁾ Most patients have a good prognosis because the tumors are usually slow growing and do not undergo malignant transformation.

References

- 1) Akhaddar A, Albouzidi A, Elmostarchid B, Gazzaz M, Boucetta M: Sudden onset of paraplegia caused by hemorrhagic spinal epidural angiolipoma. A case report. *Eur Spine J* 17 Suppl 2: S296-298, 2008
- 2) Akhaddar A, Gazzaz M, Derraz S, Rifi L, Amarti A, Aghzadi A, El Ouahabi A, El Khamlihi A: [Spinal epidural angiolipomas: a rare cause of spinal cord compression. A report of 8 cases and review of the literature]. *Neurochirurgie* 46: 523-533, 2000 (French)
- 3) Alter M: Statistical aspect of spinal cord tumor, in Vinken PJ, Bruyn GW (eds): *Handbook of Clinical Neurology*. New York, NY, North Holland Publishing Co, 1975, pp 1-22
- 4) Andaluz N, Balko G, Bui H, Zuccarello M: Angiolipomas of the central nervous system. *J Neurooncol* 49: 219-230, 2000
- 5) Anson JA, Cybulski GR, Reyes M: Spinal extradural angiolipoma: a report of two cases and review of the literature. *Surg Neurol* 34: 173-178, 1990
- 6) Bender JL, Van Landingham JH, Manno NJ: Epidural lipoma producing spinal cord compression. Report of two cases. *J Neurosurg* 41: 100-103, 1974
- 7) Berenbruch K: Ein Fall von multiplen Angiolipomen kombiniert mit einem Angiom des Rückenmark [dissertation]. Tübingen, Germany, Medical Faculty Universität Tübingen, 1890 (German)
- 8) Bucy PC, Ritchey H: Klippel-Feil's syndrome associated with compression of the spinal cord by an extradural hemangiolipoma. *J Neurosurg* 4: 476-481, 1947
- 9) Ehni G, Love JG: Intraspinal lipomas. Report of cases; review of the literature, and clinical and pathologic study [abstract]. *J Nerv Ment Dis* 102: 520-521, 1945
- 10) Fournay DR, Tong KA, Macaulay RJ, Griebel RW: Spinal angiolipoma. *Can J Neurol Sci* 28: 82-88, 2001
- 11) Francois P, Zemmoura I, Fouquet AM, Jan M, Velut S: Lateral sellar angiolipoma: a tumor illustrative of the extradural compartment of the neural axis. *J Neurosurg* 113: 1053-1058, 2010
- 12) Gelabert-Gonzalez M, Garcia-Allut A: Spinal extradural angiolipoma: report of two cases and review of the literature. *Eur Spine J* 18: 324-335, 2009

- 13) Gonzalez-Crussi F, Enneking WF, Aream VM: Infiltrating angiolipoma. *J Bone Joint Surg Am* 48: 1111–1124, 1966
- 14) Goyal RN: Epidural lipoma causing compression of the spinal cord. *Surg Neurol* 14: 77–79, 1980
- 15) Hanakita J, Koyama T: [Spinal epidural hemangiolipoma—a case report (author's transl)]. *No Shinkei Geka* 10: 313–316, 1982 (Japanese)
- 16) Henry P, Vital C, Vallat JM, Briole G, Loiseau P: [Anatomoclinical study of an extradural angiolipoma associated with an intramedullary glioblastoma]. *Rev Neurol (Paris)* 124: 385–391, 1971 (French)
- 17) Kujas M, Lopes M, Lalam TF, Fohanno D, Poirier J: Infiltrating extradural spinal angiolipoma. *Clin Neuropathol* 18: 93–98, 1999
- 18) Labram EK, el-Shunnar K, Hilton DA, Robertson NJ: Revisited: spinal angiolipoma—three additional cases. *Br J Neurosurg* 13: 25–29, 1999
- 19) Lach B, Lesiuk H: Intracranial suprasellar angiolipoma: ultrastructural and immunohistochemical features. *Neurosurgery* 34: 163–167, 1994
- 20) Leu NH, Chen CY, Shy CG, Lu CY, Wu CS, Chen DC, Guo TH: MR imaging of an infiltrating spinal epidural angiolipoma. *AJNR Am J Neuroradiol* 24: 1008–1011, 2003
- 21) Lin JJ, Lin F: Two entities in angiolipoma. A study of 459 cases of lipoma with review of literature on infiltrating angiolipoma. *Cancer* 34: 720–727, 1974
- 22) O'Donovan NA, Naik K, Maloney WJ, Llewellyn CG: Spinal angiolipoma mimicking extradural lipomatosis. *Can Assoc Radiol J* 47: 51–53, 1996
- 23) Pagni CA, Canavero S: Spinal epidural angiolipoma: rare or unreported? *Neurosurgery* 31: 758–764, 1992
- 24) Petrella G, Tamburrini G, Lauriola L, Di Rocco C: Spinal epidural angiolipoma complicated by an intratumoral abscess. Case report. *J Neurosurg* 103: 166–169, 2005
- 25) Provenzale JM, McLendon RE: Spinal angiolipomas: MR features. *AJNR Am J Neuroradiol* 17: 713–719, 1996
- 26) Rabin D, Hon BA, Pelz DM, Ang LC, Lee DH, Duggal N: Infiltrating spinal angiolipoma: a case report and review of the literature. *J Spinal Disord Tech* 17: 456–461, 2004
- 27) Sciort R, Akerman M, Dal Cin P, De Wever I, Fletcher CD, Mandahl N, Mertens F, Mitelman F, Rosai J, Rydholm A, Tallini G, Van den Berghe H, Vanni R, Willen H: Cytogenetic analysis of subcutaneous angiolipoma: further evidence supporting its difference from ordinary pure lipomas: a report of the CHAMP Study Group. *Am J Surg Pathol* 21: 441–444, 1997
- 28) Trabulo A, Cerqueira L, Monteiro J, Roque P, Reis FC, Coelho MR: Spinal angiolipomas revisited: two case reports. *Acta Neurochir (Wien)* 138: 1311–1319, 1996
- 29) Turgut M: Spinal angiolipomas: report of a case and review of the cases published since the discovery of the tumour in 1890. *Br J Neurosurg* 13: 30–40, 1999
- 30) Yamashita K, Fuji T, Nakai T, Hamada H, Kotoh K: Extradural spinal angiolipoma: report of a case studied with MRI. *Surg Neurol* 39: 49–52, 1993

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