

Exploring a Tumor Spectrum in a Patient with Familial Angiolipomatosis

Abstract

Angiolipomas are uncommon spinal tumors which differ from their cutaneous counterparts in having larger caliber vascular stroma. Although slow growing, they can cause rapid spinal cord compression and sudden-onset sensorimotor symptoms due to vascular engorgement, hemorrhage, or thrombosis. The goal of surgery is spinal decompression, and favorable outcome is the rule. We report a patient with spinal angiolipoma, vertebral hemangioma along with subcutaneous lipomas and angiolipomas, exhibiting the entire histopathological spectrum of these related soft-tissue tumors. Analysis of his family tree revealed a hereditary predilection. Familial angiolipomatosis is an uncommon genetic condition which has not been reported to occur with spinal angiolipomas thus far.

Keywords: *Angiolipoma, familial angiolipomatosis, spinal angiolipoma*

Introduction

Lipomas, angiolipomas, and hemangiomas represent a continuous histopathological spectrum of related tumors.^[1] These tumors are benign and slow growing in nature. The main differentiating factor between them is the ratio between vascular and adipose components. The vascular fraction comprises <10% of tumor mass in lipomas, 15%–40% in angiolipomas,^[2] whereas hemangiomas consist almost entirely of malformed vasculature with scanty intervening stroma.

Lipomas and angiolipomas occur sporadically, but have been found to have a familial incidence as well. Familial angiolipomatosis is usually included within the broader spectrum of familial lipomatosis, mirroring the classification of the tumors they encompass.^[3]

While an overwhelming majority of hemangiomas are sporadic, an autosomal dominant pattern has been described.^[4] In spite of similarities between lipomas, angiolipomas, and hemangiomas, individual cases where these three histological types occur together are uncommon and as per our literature review have never been reported.

We describe a patient of familial angiolipomatosis who presented with a dorsal extradural angiolipoma, with

asymptomatic multiple subcutaneous lipomas, angiolipomas, and a large lumbar vertebral hemangioma, representing the spectrum of histopathologically related soft-tissue tumors within the same patient.

Case Report

A 32-year-old man presented with a 20-day history of progressively severe imbalance on walking with bilateral lower limb weakness, gradually increasing numbness extending from the upper abdomen to both lower limbs, and dysesthesia on walking. He also complained of straining at micturition with a weak urinary stream over the last 20 days. On inquiry, he had multiple painless soft-tissue swellings over his left forearm and abdomen, with a family history of similar swellings in his father and two brothers.

On general examination, his swellings were well defined, nonfluctuant, and not associated with tenderness, pruritis, or overlying skin changes. On neurological examination, there was increased tone with Grade 4 power in both lower limbs. Hyperreflexia was elicited in bilateral lower limbs with ill-sustained ankle clonus, bilateral Babinski's sign, and absent abdominal reflex. There was partial sensory loss for touch and pinprick below D6 levels and impaired proprioception in both lower limbs.

Spinal magnetic resonance imaging (MRI) demonstrated a large extradural lesion

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extending from D5 to D8 levels causing severe compression of the cord. The lesion was hyperintense on both T1-weighted imaging (T1WI) and T2WI, with multiple flow voids within the lesion. On fat suppression images, the lesion showed suppression suggesting fatty component in the lesion; however, there was intense contrast enhancement suggestive of spinal epidural angioliipoma or spinal arteriovenous malformations (AVM). A hemangioma in L1 vertebral body was also discovered incidentally in the imaging [Figures 1 and 2].

A spinal digital subtraction angiography (DSA) confirmed the diagnosis of an angioliipoma with multiple vessels feeding the lesion from right D5 and bilateral D6 and D7 intercostal arteries. There was no early venous drainage, hence ruling out the possibility of an AVM, while supporting the diagnosis of an angioliipoma.

Ultrasonography of the forearm swellings revealed three ovoid, well-defined, hyperechoic lesions within the ventral forearm space suggestive of lipomas. A Doppler scan revealed venous flow within peripherally arranged vessels without evidence of intralesional flow in some lesions, most likely representing angioliipoma. A histopathological diagnosis could not be obtained to confirm this, as the swelling was not excised.

Embolization of the spinal lesion was deferred in view of the diffuse nature of the feeders. Standard D5–D8 laminectomy was performed. A reddish yellow tumor was identified in the epidural region merging with the normal epidural fat at both the edges. It was friable, soft, and extremely vascular with interspersed spongy and hemorrhagic areas. Vascular supply of the lesion consisted of multiple large epidural feeders which made hemostasis challenging. The tumor was slowly devascularized by coagulating the intercostal feeders and was radically excised to achieve decompression of the cord. Complete excision of the tumor was achieved, which was confirmed on postoperative MRI. Histopathological examination of the excised mass revealed fibrofatty tissue embedded within numerous dilated vessels, lined by single flattened epithelium along with few areas of hyalinization, consistent with an angioliipoma.

The patient had immediate postoperative improvement of his weakness, dysesthesia, and urinary complaints and was mobilized on the 2nd postoperative day.

At 6 months' follow-up examination, the patient had Grade 5 power in both lower limbs and complete recovery of sensory deficits. There were no urinary or bowel complaints. At 1-year follow-up, there were no major complaints; however, the multiple subcutaneous swellings had increased in size and number.

Discussion

Angioliipomas are considered a histological subtype of lipomas, accounting for 5%–17% of all lipomatous lesions.^[5] These are benign, slow-growing lesions composed of mature adipocytes admixed with abnormal blood vessels, which are usually of capillary size.^[6] Multiple lipomatous lesions in individuals are more likely to be angioliipomas, whereas isolated lesions are most commonly simple lipomas.^[4]

Spinal angioliipoma is an extremely scarce entity constituting 0.04%–1.2% of all spinal tumors,^[7] with just over 100 cases reported in literature.^[8] However, its clinical and pathological features lean toward hemangiomatous tumors, meriting its distinction from cutaneous angioliipomas.^[6]

They are most commonly located at the dorsal and lower cervical region along the midline, conforming to the last site of closure of embryonic neural arch at its rostral end.^[9] These lesions commonly present as sudden-onset weakness and sensory loss in the lower limbs.^[10] The rapid symptom progression may be attributed to vascular engorgement, intralesional hemorrhage, sudden thrombosis, change in capillary caliber, or vascular steal phenomenon.^[7,8,10-13] Vascular engorgement may have been the most likely culprit in our case which was characterized by significant intraoperative blood loss during tumor removal.

Like simple lipomas, spinal angioliipomas are hyperintense on both T1WIs and T2WIs.^[14] Provezale and McLendon reported that large hypointense foci within spinal angioliipomas on noncontrast T1WIs are correlated with increased vascularity and most lesions enhance with gadolinium administration, whereas T2WI can be variable, but are commonly hyperintense.^[15,16] Thus, a conspicuous differentiating feature of angioliipomas on MRI is an intense contrast enhancement which is not seen with lipomas.

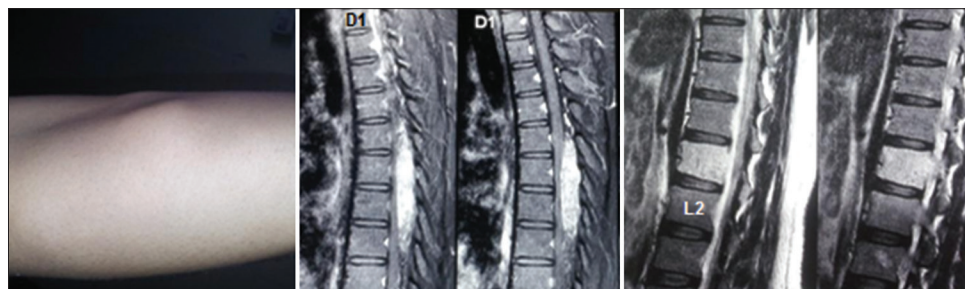


Figure 1: Spectrum of lesions seen in this patient. From left to right: Subcutaneous angioliipoma, dorsal epidural angioliipoma, and vertebral hemangioma



Figure 2: Sagittal and axial cuts of magnetic resonance imaging showing large, extradural, contrast-enhancing lesion extending from D5 to D8 with hemangioma in L1 vertebral body. (a) T1-weighted images. (b) Fat-suppressed postcontrast image. (c) T2-weighted images

DSA helps in delineating the vascular supply and in differentiating spinal angioliomatosis from AVMs. Embolization is not indicated as the feeders are diffuse and may jeopardize the normal supply of the cord. Total surgical excision is considered the best line of management in these cases.^[17]

Spinal angioliomatosis have a significantly higher caliber of vascular component in comparison with their cutaneous counterparts and therefore, may even be considered a distinct entity.^[6,18]

Familial multiple angioliomatosis is a rare and benign condition which is transmitted in an autosomal recessive fashion, although certain cases of autosomal dominant inheritance have been noted.^[19] It has been traditionally included within the broader definition of familial multiple lipomatosis.^[3] The patient at hand is among the second generation within the family to be affected. No third-generation family members are symptomatic thus far, the oldest member being 14 years of age. However, one must note that most cutaneous angioliomatosis arise after the second decade of life, and one cannot rule out the possibility of third-generation family members also suffering from the condition [Figure 3].

Hemangiomas and lipomas are two ends of a continuous histopathological spectrum, where angioliomatosis form the intermediate variety. Lipomas are tumors of mature adipocytes without cellular atypia and are the most common soft-tissue tumors of adulthood.^[20] Angioliomatosis are considered a rarer subtype of lipomas where mature

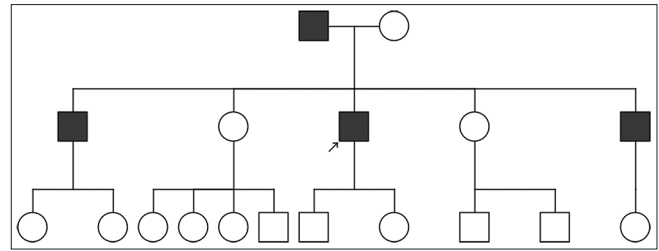


Figure 3: Pedigree chart of the family

adipocytes are dispersed within a prominent vascular stroma. Hemangiomas are endothelial tumors consisting of malformed blood vessels. The more vascular, invasive, and aggressive spinal angioliomatosis would represent a shift toward the hemangioma end of the spectrum. Despite their histopathological correlation and their common mesenchymal origin, these tumors have markedly varied pathogenesis not only between themselves, but also between subtypes of the same tumor. Notably, genetic aberrations seen in cutaneous lipomas are not present in angioliomatosis.^[21] Given their mottled pathologic origin, the simultaneous occurrence of multiple tumor types within the same patient seems striking, more so when considering the familial nature of his affliction. There is only one case reported in literature which describes the occurrence of spinal angioliomatosis in a case of familial lipomatosis.^[22] Our patient is unique in that he probably represents a case of familial angioliomatosis with multiple subcutaneous lesions, presenting with an associated dorsal angioliomatosis as well as a lumbar vertebral hemangioma.

Conclusion

While lipomas, angioliomatosis, and hemangiomas represent a continuous histological spectrum of mesenchymal tumors, they differ greatly in their pathogenesis. Spinal angioliomatosis are uncommon neoplasms, the origin of which is uncertain. Although they are considered conspicuously dissimilar to their cutaneous counterparts, we report the coincidence of both spinal and cutaneous lesions in this case of familial angioliomatosis with lumbar vertebral hemangioma. Their coexistence in a singular patient with close histopathological features makes us hypothesize that the three lesions are nothing but natural progression of a single tumor type - either a hemangioma’s vascular component gradually regresses to form an angioliomatosis and eventually lipoma or a lipoma acquires progressive vascularity to reverse the aforementioned trend.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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