

Short Commentary

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The Sinewy Swirls- Angioleiomyoma

Anubha Bajaj*

Histopathologist in A.B. Diagnostics, New Delhi, India.

***Corresponding Author: Anubha Bajaj**

Histopathologist in A.B. Diagnostics, New Delhi, India

Email: anubha.bajaj@gmail.com

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Abstract

Angioleiomyoma is an exceptional, benign, solitary neoplasm originating from tunica media of vascular smooth muscle. The neoplasm commonly represents as a uniform, well defined, solitary, soft, nodular, gradually progressive, bluish or purple lesion commonly beneath <2 centimetres magnitude with an absence of accompanying inflammation. Tumefaction is composed of thickened smooth muscle fibres and vascular channels with circumscribing attenuated fibrous tissue capsule. Additionally, the lesion may denominate organizing thrombus, mature adipose tissue cell aggregates and lymphocytic infiltrate. The neoplasm is immune reactive to desmin, myosin, α -smooth muscle actin, HHF35, calponin and h-caldesmon. Angioleiomyoma necessitates a segregation from angiomyolipoma, leiomyoma with bizarre nuclei, glioblastoma multiforme, arteriovenous malformation, cavernous haemangioma, angiofibroma, primary intracranial myopericytoma, glomus tumour, traumatic neuroma, eccrine spiradenoma, angiolipoma, ganglion cyst, neuroma, fibroma, lipoma, neurofibroma, neurilemmoma, salivary gland neoplasms as mucocoele, pleomorphic adenoma, vascular neoplasms as lymphangioma, haemangioma, pyogenic granuloma or dermoid cyst. Comprehensive surgical excision of the lesion with a tumour -free perimeter is an optimal treatment strategy.

Keywords: Angioleiomyoma, Neoplasm.

Short commentary

Leiomyoma is subdivided into pertinent categories designated as angioleiomyoma and leiomyoma arising from deep-seated soft tissue. Angioleiomyoma is an exceptional, benign, solitary neoplasm which originates from tunica media of vascular smooth muscle. Tumefaction is composed of vascular articulations with thickened smooth muscle fibres and is devoid of elastic tissue. The essentially benign, painful, subcutaneous or deep-seated, dermal

neoplasm comprised of vascular configurations and smooth muscle emerges as a morphological continuum to myopericytoma and myofibroma [1]. Angioleiomyoma is additionally designated as an “angiomyoma” or a “vascular leiomyoma”. The neoplasm is usually detected and appropriately categorized upon comprehensive surgical excision [1].

Disease characteristics: As estimated by World Health Organization, angioleiomyoma is a frequently discerned variant

of leiomyoma with an incidence of around 74%. Nearly 55.9% instances of the vascular neoplasm appear reddish, bluish or purple whereas roughly 44.1% tumours are grey, white or mucosa-tinged [1,2]. Angioleiomyoma is generally situated within the lower and upper extremities, face or oral cavity although no site of tumour emergence is exempt.

The neoplasm is commonly discerned within middle aged individuals between 30 years to 60 years. Males are incriminated at a median age of 53 years whereas females are implicated at a median age of 38 years [2,3].

Majority of tumefaction are miniature with a tumour diameter below <2.0 centimetres although neoplasms vary from 0.5 centimetres to 2.0 centimetres in magnitude with median dimension of 1.0 centimetre. Symptomatic individuals depict a median tumour magnitude of 1.0 centimetre whereas asymptomatic tumefaction demonstrates a median tumour diameter of 0.7 centimetres [2,3]. Angioleiomyoma is associated with a female predominance, is commonly discerned within lower extremities of female subjects and head and neck or upper extremities in male subjects [2,3]. Malignant metamorphoses is exceptional, emerges as a primary, vascular leiomyosarcoma or malignant tumour reoccurrence of a pre-existing angioleiomyoma and accounts for around 7% of soft tissue sarcomas [2,3].

Clinical elucidation: Angioleiomyoma is a uniform, well defined, solitary, soft, nodular, gradually progressive, bluish or purple lesion commonly beneath <2 centimetres magnitude with an absence of accompanying inflammation. Tumefaction is mobile, tender and demonstrates an elevated superimposed layer of stratified squamous epithelium. Alternatively, an exophytic, nodular, reddish-purple, well circumscribed lesion of varying magnitude with unremarkable superimposed mucosa can be] discerned [3,5]. The neoplasm manifests as a painful or painless nodule. Concurrent pain is described as paroxysmal, drilling or radiating. Certain centric lesions of oral angioleiomyoma may be painful. Neoplasms confined to the facial region or head are commonly painless [3].

The classic clinical symptom of angioleiomyoma is a painful nodule occurring in association with or absence of tenderness which is discerned in an estimated 45% to 70% individuals. Pain appears on account of stretching of nerves entrapped within the tumour parenchyma or tumour capsule or originates due to efflux of mast cell mediators.

Pain can be paroxysmal and is precipitated by fleeting touch, exposure to wind, cold, menses, variations in extraneous pressure or imperceptible stimuli [3,5]. Morimoto in 1973 categorized angioleiomyoma into three distinctive subtypes denominated as

- solid or capillary subtype which is painful, appears in an estimated 66% neoplasms and arises within lower extremities, generally within female subjects [4].
- venous subtype which is painless and is discerned within males wherein around 23% lesions commonly arise within the head and neck [4].

- cavernous subtype is exhibited in roughly 11% instances where in tumours are common within the upper extremity or head [4]. Angioleiomyoma can delineate an amalgamation of solid, venous or cavernous subcategories. Nevertheless, the subtypes are devoid of pertinent clinical significance. Also, clinical categories do not appear to be concordant with histological subtypes [4]. Angioleiomyoma confined to the hand is minimally painful and of venous or cavernous subtype. Pain is engendered due to localized tissue ischemia arising from contraction of blood vessels feeding the neoplasm or mediated through nerve fibres traversing tumour parenchyma. Miniature nerves fibres are observed entrapped within the tumour capsule or tumour stroma [5].

Histological elucidation: Macroscopically, the neoplasm is irregular, lobulated, smooth, spherical, pearly grey and well encapsulated. Tumefaction can emerge as a well demarcated, homogenous nodule confined to deep-seated dermis or arise as a firm, sharply circumscribed, grey/white or brown nodule of magnitude below <2 centimetres. Cut surface is glistening, grey/white or reddish brown on account of enlarged number of blood vessels [5,6]. On fine needle aspiration cytology, moderate or sparsely cellular smears are composed of uniform, spindle-shaped cells admixed with variable proportion of smooth muscle cells and fragments of collagenous tissue. Macrophages, mature adipose tissue cells or enlarged, ganglion-like cells are occasionally discerned [5,6]. On microscopy, tumour nodule is comprised of thickened smooth muscle fibres and vascular channels circumscribed by an attenuated fibrous tissue capsule. Although constituted by vascular configurations of varying magnitude admixed with smooth muscle fascicles, lesions may denominate organizing thrombus, mature adipose tissue cell aggregates and lymphocytic infiltrate. Miniature foci of hyaline or myxoid alterations are observed [5,6]. The neoplasm exhibits enlarged, vascular channels of diverging calibre encompassed with thickened walls along with irregular dissemination of spindle-shaped cells. Spindle-shaped tumour cells are imbued with elongated, blunt-ended nuclei and eosinophilic cytoplasm. Thereby, smooth muscle bundles are configured which are admixed with interlacing collagen fibres and distended, slit-like vascular articulations.

Smooth muscle fibres configure whorled fascicles adherent to the vessel wall. Atypical mitosis and cellular or nuclear pleomorphism are absent. Well circumscribed fascicles of mature smooth muscle cells are encompassed with vascular lumen layered with normal endothelium and an absence of elastic lamina [5,6].

- solid subtype is composed of compact, closely aggregated fascicles of smooth muscle.
- venous subtype is comprised of vascular articulations with thick, muscular walls which intermingle with smooth muscle bundles.
- cavernous subtype is constituted by distended vascular channels with minimal quantities of smooth muscle which commingle with adjacent smooth muscle fascicles [4]. The neoplasm may exhibit foci of cartilaginous or adipose

tissue metaplasia or demonstrate bizarre, degenerative cells recapitulating a symplasmic uterine leiomyoma. Prominent tumour calcification within acral sites is exceptional. Scattered epithelioid cells or epithelioid cell granuloma is infrequently observed. Tumour necrosis, haemorrhage, mitotic activity, features of vasculitis or fibromuscular dysplasia are absent [5,6].

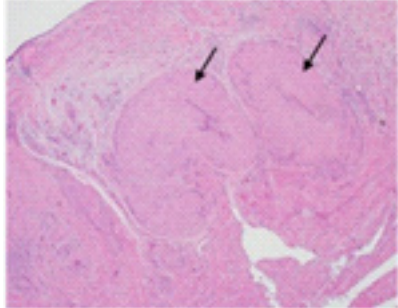


Figure 1: Angioleiomyoma delineating vascular configurations with a significant smooth muscle component and an absence of elastic layer [10].

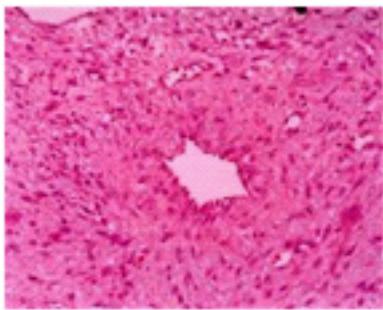


Figure 2: Angioleiomyoma enunciating endothelium coated vascular articulations with an extensive smooth muscle cell layer and an absence of elastic lamina [11].

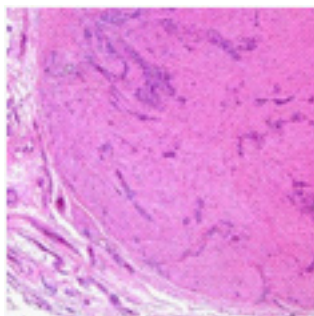


Figure 3: Angioleiomyoma depicting vascular articulations constituted by expansive smooth muscle bundles, a lack of elastic layer and a prominent adventitial layer [12].

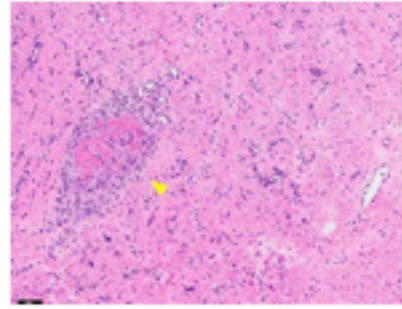


Figure 4: Angioleiomyoma exemplifying vascular channels composed of extensive smooth muscles and an absence of elastic layer [13].

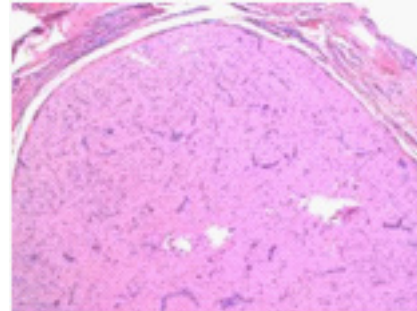


Figure 5: Angioleiomyoma exhibiting vascular arrangements with hypertrophic smooth muscle, prominent tunica media and an absence of elastic layer [14].

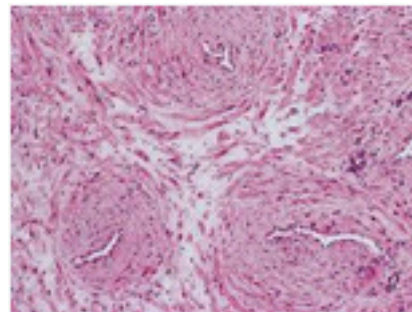


Figure 6: Angioleiomyoma delineating endothelium layered vascular configurations with hypertrophied smooth muscle layer and an absence of elastic lamina [15].

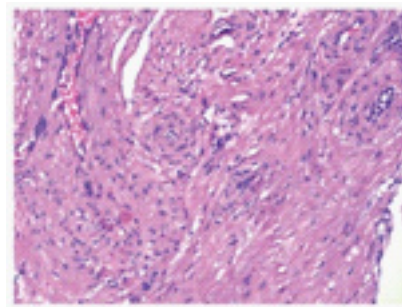


Figure 7: Angioleiomyoma depicting endothelial layered vascular articulations with smooth muscle cells and an absence of elastic lamina [16].

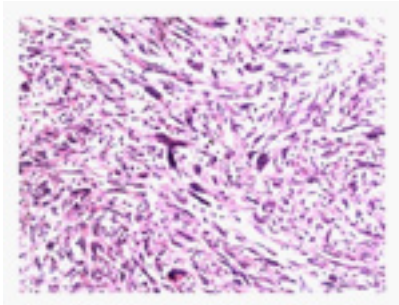


Figure 8: Angioleiomyoma demonstrating endothelium layered vascular channels associated with cellular atypia of the surrounding stroma and incorporated hyperchromatic nuclei [17].

Immune histochemical elucidation: Smooth muscle cells are immune reactive to α - smooth muscle actin (α -SMA) in around 90% tumefaction. The neoplasm is immune reactive to desmin, myosin, HHF35, calponin and h-caldesmon. Tumour cells can be highlighted with trichrome stains. Miniature nerve fibres are immune reactive to vimentin, type IV collagen and S100 protein. Endothelial cells are immune reactive to CD34. Nasal lesions are variably immune reactive to progesterone receptors. Proliferative index Ki67 or MIB-1 is minimal [2,3]. The neoplasm is immune non reactive to human melanoma black 45 (HMB-45) antigen, oestrogen receptors or glial fibrillary acidic protein (GFAP) [6,8].

Differential Diagnosis: Angioleiomyoma necessitates a segregation from angiomyolipoma or leiomyoma with bizarre nuclei which can be differentiated as

- angiomyolipoma demonstrates a prominent component of mature adipose tissue and is immune reactive to human melanoma black 45 (HMB45) antigen [7].
- leiomyoma with bizarre nuclei is devoid of a prominent vascular component [7]. Intracranial angioleiomyoma requires demarcation from diverse entities such as glioblastoma multiforme (GBM), arteriovenous malformation (AV), cavernous haemangioma, angiofibroma (AFM) and myopericytoma (MPC) [8].
- glioblastoma multiforme is an infiltrating, high grade neoplasm which is a cystic lesion associated with tumour necrosis, enhanced tumour metabolism, prominent mitosis and capacity for proliferation. Diffuse vascular articulations are absent. The neoplasm is immune reactive to glial fibrillary acidic protein (GFAP) [8].
- arteriovenous malformation (AVM) is an anomaly comprised of tangled clusters of tortuous arteries, veins and atypical, interconnected vascular channels [8].
- cavernous haemangioma is composed of peripheral vascular channels or an enlarged blood filled sinus [8].
- angiofibroma is frequently denominated as a juvenile nasopharyngeal angiofibroma with accompanying intracranial extension. The neoplasm is commonly discerned within teenage males. Juvenile nasal angiofibroma demonstrates an abundant vascular component in the absence of smooth muscle component. The neoplasm is immune reactive to CD34 and is immune

non reactive to smooth muscle actin (SMA) [8].

- primary intracranial myopericytoma (MPC) is an exceptional neoplasm which morphologically recapitulates angioleiomyoma as the dual conditions are constituted by myoid- appearing cellular component with diffuse vascular articulations. However, myopericytoma depicts elliptical to spindle- shaped cells which concentrically circumscribe perivascular zones. The neoplasm is immune non reactive to desmin [8].

Angioleiomyoma emerging as a painful nodule requires segregation from glomus tumour, traumatic neuroma, eccrine spiradenoma or angiolipoma. Tumours confined to the extremity mandate distinction from a ganglion cyst, neuroma, fibroma, lipoma, neurofibroma, neurilemmoma or an inclusion cyst [7,9]. Angioleiomyoma requires a clinical demarcation from mesenchymal neoplasms such as leiomyosarcoma, salivary gland neoplasms as the mucocoele or pleomorphic adenoma, vascular neoplasms as lymphangioma, haemangioma or pyogenic granuloma along with soft tissue cysts as dermoid cyst [7,9]. Angioleiomyoma situated within the hard palate or lesions adjacent to the teeth necessitate distinction from a periodontal lesion [7,9].

Investigative assay: Ultrasonography depicts a well defined, minimally echoic, hyper-vascular tumour nodule with a distinctive tumour perimeter and homogenous intrinsic architecture, indicative of a benign neoplasm. Magnetic resonance imaging (MRI) features are non specific. T2 weighted imaging delineates a mass with mixed foci of hyper-intense and iso- intense regions in concordance with skeletal muscle with a hypo-intense tumour periphery [7,9]. Tumour discernment is adequately achieved with surgical excision and a cogent histological examination [10].

Therapeutic options comprehensive surgical excision of the lesion is an optimal treatment strategy. Tumour associated morbidity is minimal. Oral angioleiomyoma is preferably subjected to surgical excision. Despite an extensive vascular component, profuse surgical haemorrhage is exceptionally encountered [9,10]. Comprehensive surgical extermination with a tumour free perimeter is recommended in order to circumvent tumour reoccurrence. Localized tumour reoccurrence is exceptional and attributed to incomplete surgical resection or encountered in deep-seated lesions [9,10].

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