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Commentary

Embroiled and Tangled-Angiofibroma of Soft Tissue

Anubha Bajaj*

Department of Histopathology, Panjab University, A.B. Diagnostics, India

***Corresponding Author:** Anubha Bajaj, Department of Histopathology, Panjab University, A.B. Diagnostics, India.

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Angiofibroma of soft tissue is a benign, well defined, fibroblastic neoplasm incriminating soft tissues of extremities. Arising in concurrence with joints or fibrotendinous structures, tumefaction is comprised of uniform, spindle shaped cells intermingled within a variably myxoid and collagenous stroma imbued with a network of thin walled, branching vascular articulations. Cytogenetically, neoplasm delineates NCOA2 genetic rearrangements.

Angiofibroma of soft tissue commonly appears within middle aged adults and demonstrates a wide age of disease emergence ranging from 6 years to 68 years. A mild female preponderance is encountered.

Typically, angiofibroma of soft tissue is confined to lower extremities although sites involving or adjacent to large joints as knee joint may be incriminated. Besides, locales such as dorsal region, abdominal wall, pelvic cavity or breast may be infrequently incriminated [1,2].

Generally, tumefaction is confined to subcutaneous region although intramuscular and deep seated lesions may be encountered.

Angiofibroma of soft tissue is posited to arise due to occurrence of near diploid karyotypes along with recurrent chromosomal translocation t(5;8)(p15;q13) with consequent fusion of AHRR gene confined to chromosome band 5p15 and NCOA2 gene confined to chromosome 8q13 [1,2].

AHRR::NCOA2 genetic chimera with basic helix - loop - helix and PAS domains confined to AHHR gene along with dual transcriptional activation domains of NCOA2 may emerge within 60% to 80% neoplasms. Additionally, variant GTF2I::NCOA2 or GAB1::ABL1 genetic fusions may be documented.

AHRR::NCOA2 genomic chimera may upregulate AHRR/ARNT genetic signalling along with emergence of aryl hydrocarbon receptor target genes [1,2].

Of obscure aetiology, angiofibroma of soft tissue demonstrates genetic rearrangement of NCOA2, genetic fusion gene AHRR::NCOA2 or a variant genomic fusion.

Besides, variant GTF2I::NCOA2 or GAB1::ABL1 genetic fusions are documented.

Also, NCOA2::ETV4, ETV4::AHRR, P4HA2::TBCK and TBCK::P4HA2 may ensue [1,2].

Angiofibroma of soft tissue appears as a gradually progressive, painless tumefaction demonstrating an extended preoperative duration. Majority of lesions emerge as sharply demarcated neoplasms [2,3].

Upon gross examination, a well demarcated, nodular or multinodular, solid tumefaction may be associated with infiltration into surrounding viscera and organs. Tumour magnitude varies from 1.2 centimetres to 10.0 centimetres with mean tumour magnitude of 5.1 centimetres.

Cut surface is white to yellow and frequently glistening. Foci of cystic or haemorrhagic degeneration may be delineated [2,3].

Upon microscopy, the well circumscribed neoplasm depicts a mildly lobulated architecture. Neoplasm is composed of alternating myxoid and collagenous areas. Tumour cellularity varies within diverse zones. Neoplasm is composed of uniform, bland, spindle shaped cells embedded within a prominent vascular network. Constituent spindle shaped cells appear imbued with inconspicuous, pale, eosinophilic cytoplasm, miniature ovoid or tapering nuclei with irregular nuclear contour, fine nuclear chromatin and indistinct nucleoli.

Generally, cytological and nuclear atypia or hyperchromatic nuclei are absent [2,3].

Encompassing vascular network is comprised of innumerable miniature, thin walled vascular articulations which appear regularly disseminated throughout the stromal parenchyma. Also, medium or enlarged vascular articulations with variably thickened walls may be discerned. Deposition of perivascular collagen and marked hyalinization or fibrinoid necrosis of walls of vascular articulations of intermediate magnitude may occur.

Occasionally, degenerative atypia of the tumefaction may be encountered. Degenerative alterations may be focal and are comprised of chronic inflammation, haemorrhage and aggregates of foamy histiocytes [2,3].

A variably intense inflammatory infiltrate comprised of lymphocytes may be delineated wherein lymphocytic invasion is preponderantly distributed within the perivascular zone [2,3].

Angiofibroma of soft tissue exhibits nuclear expression of NCOA2 gene. Besides, variable expression of epithelial membrane antigen (EMA), CD34 or smooth muscle actin (SMA) may ensue. Occasionally, dendritic cells exhibit desmin.

Angiofibroma of soft tissue is immune non reactive to S100 protein, cytokeratin CK AE1/AE3, MNF116 or STAT6 [4,5].

Angiofibroma of soft tissue requires segregation from neoplasms as cellular angiofibroma, solitary fibrous tumour, low grade fibromyxoid sarcoma, low grade myxofibrosarcoma or myxoid liposarcoma.

Appropriate detection of angiofibroma of soft tissue requires cogent interpretation of clinical features, radiological findings and histopathological manifestations [4,5].

Upon imaging, neoplastic infiltration into adjacent anatomic structures or viscera may be encountered [4,5].

Magnetic resonance imaging (MRI) demonstrates a well circumscribed tumefaction. Characteristically, imaging signals and image enhancement appears variable and contingent to cellular, myxoid, collagenous and vascular constituents of the neoplasm [4,5].

Upon T1 weighted magnetic resonance imaging, an isointense signal intensity roughly equivalent to skeletal muscle is encountered. Upon T2 weighted magnetic resonance imaging, a heterogeneously elevated signal intensity is encountered [4,5].

Tumour	Clinical features	Histological feature	IHC	Molecular features	Clinical behaviour
Angiofibroma of soft tissue	Subcutaneous soft tissue of extremities involving or adjacent to large joints	Uniform spindle cells alternating with myxoid and collagenous stroma and network of innumerable, thin walled, branching vessels	Variable expression of CD34, EMA, SMA,	AHRR-NCOA2 fusion (~80%), rarely GTF2I-NCOA2 or GAB1-ABL1 fusions	Benign. Local recurrence rare. Metastatic potential absent
Cellular angiofibroma	Superficial soft tissues of vulvovaginal region	Uniform short spindle cells in an oedematous, fibrous stroma with short bundles of delicate collagen and numerous small to medium, thick walled blood vessels with round or branching lumens	CD34, loss of Rb expression of tumour cells	Chromosomal 13q14 (RB1) deletion	Benign. Recurrence or metastasis ensues with sarcomatous change
Solitary fibrous tumour	Pleura and extrapleural location	Haphazardly arranged uniform spindle to ovoid cells within a variably collagenous stroma admixed with branching, stag-horn blood vessels with perivascular hyalinization	STAT6 (nuclear), CD34	NAB2-STAT6 fusion	Variable. Lesion classified as low, intermediate, high risk contingent to age, mitosis, tumour magnitude, necrosis.
Low grade fibromyxoid sarcoma	Deep soft tissue of proximal extremities and trunk of young adults. Tumour demonstrates a long latent period	Bland spindle to plump cells with short fascicles or whirling pattern in an alternating hypocellular collagenous or cellular myxoid stroma with arcades of small vessels	MUC4, EMA, SMA	FUS-CREB3L2 fusion (~90%), FUS-CREB3L1 and EWSR1-CREB3L1 fusions (uncommon)	Low grade malignant with delayed reoccurrence or metastasis
Myxoid liposarcoma	Deep soft tissue of extremities, commonly thigh	Uniform, round to ovoid cells with variable small lipoblasts within a myxoid stroma with branching capillary vasculature	NY-ESO-1, S100 protein,	FUS-DDIT3 fusion (> 95%), EWSR1-DDIT3 genetic fusion (3%).	High grade malignant. Local reoccurrence (25%), distant metastasis (~60%).
Low grade myxofibrosarcoma	Subcutaneous tissue of limbs in elderly subjects	Multinodular, infiltrative, hypocellular, atypical fibroblastic cells and pseudo-lipoblasts in a prominent myxoid stroma with curvilinear blood vessels.	CD34 and SMA	N/A	Low grade malignant, local reoccurrence (~40%. Low risk of metastasis

Table: *Differential diagnosis of angiofibroma of soft tissue [2,3].*

IHC: Immunohistochemistry; SMA: Smooth Muscle Actin; EMA: Epithelial Membrane Antigen.

Upon T1 weighted imaging with gadolinium contrast (T1C+Gd), a variable, homogeneous signal intensity with peripheral image enhancement may be encountered.

Angiofibroma of soft tissue can be appropriately subjected to conservative therapy with simple surgical extermination of the neoplasm. Angiofibroma of soft tissue demonstrates a benign clinical course. Localized tumour reoccurrence is exceptionally discerned whereas distant tumour metastasis remains undocumented [4,5].

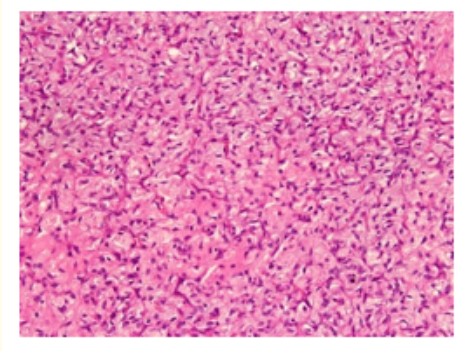

Figure 1: *Angiofibroma of soft tissue demonstrating uniform population of spindle shaped cells enmeshed within alternating zones of myxoid and collagenous stroma imbued with a network of innumerable, thin walled vascular articulations [6].*

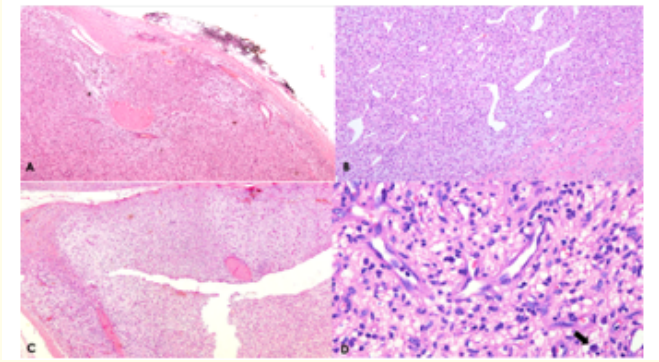

Figure 2: *Angiofibroma of soft tissue delineating aggregates of uniform spindle shaped cells encompassed within alternating zones of myxoid or collagenous stroma incorporated with a network of innumerable, thin walled vascular configurations [7].*

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6. Image 1 Courtesy: Pathology outlines.

7. Image 2 Courtesy: Cureus.com.

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