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Short Communication

The Lardaceous Sinew-Angiomyolipoma

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Angiomyolipoma represents as an exceptionally discerned, benign, mesenchymal neoplasm constituted of an admixture of smooth muscle, mature adipose tissue and thickened, dysmorphic vascular articulations. Angiomyolipoma configures as a member of perivascular epithelioid cell (PEComa) tumour family.

Classic variant of angiomyolipoma manifests with variable quantification of diverse mesenchymal components. Epithelioid variant of angiomyolipoma exhibits a definite potential for distant metastasis. Certain neoplasms may represent with significant sclerosis.

Angiomyolipoma may arise within renal parenchyma or divergent extra renal sites. Angiomyolipoma is generally asymptomatic. Tumefaction may be incidentally discovered in individuals subjected to imaging for various disorders.

Majority (~80%) of neoplasms exhibit sporadic disease occurrence. Alternatively, angiomyolipoma may concur with tuberous sclerosis.

Angiomyolipoma is commonly discerned within middle aged or adult subjects. A female preponderance is encountered.

Neoplasm may concur with tuberous sclerosis wherein the tumefaction incriminates young subjects and is devoid of specific gender predilection [1,2].

Angiomyolipoma commonly involves renal parenchyma and constitutes an estimated ~1% of renal neoplasms. Besides, tumefaction may arise within diverse extra renal sites as hepatic parenchyma or regional lymph nodes. Ovarian stroma may be exceptionally implicated.

Classic angiomyolipoma frequently exhibits genomic mutations of TSC2 gene. Sporadic neoplasms delineate mutations within TSC1 gene. Chromosome 16p enunciates copy neutral loss of heterozygosity (LOH). Genetic mutation may be discerned within chromosome 5q.

Epithelioid angiomyolipoma exemplifies mutation within p53 gene. Exceptionally, 'fat predominant' angiomyolipomas demonstrate MDM2 genetic amplification, as discerned by fluorescent in situ hybridization(FISH) [1,2].

Angiomyolipoma is possibly engendered from perivascular epithelioid cells, a cellular component devoid of specific visceral counterpart. Generally asymptomatic, miniature neoplasms may concur with tuberous sclerosis and appear discernible upon cogent disease screening.

Additionally, individuals devoid of tuberous sclerosis may delineate concurrence of angiomyolipoma with renal cell carcinoma, especially clear cell variant [2,3].

Angiomyolipoma may configure enlarged, multifocal neoplasms which may extend into renal vein or inferior vena cava.

Enlarged lesions of the benign, classic angiolipoma may be associated with tumour haemorrhage.

Upon cytological examination, elliptical to spindle shaped cells and cohesive stromal fragments appear commingled with foci of mature adipose tissue and branching vascular articulations. Cellular component is encompassed within a haemorrhagic background. Mitotic figures are absent [2,3].

Upon gross examination, unilateral, uni-focal, well circumscribed, non encapsulated tumour with a 'pushing' perimeter is encountered. Tumour magnitude varies from 0.5 centimetres to 25 centimetres with mean tumour diameter of 6 centimetres. Cut surface exhibits a reddish vascular component, grey/white smooth muscle component or yellowish adipose tissue component. Exceptionally, neoplasm may depict a cystic component.

The essentially benign angiomyolipoma may incriminate intrarenal venous system, renal vein or inferior vena cava. Around ~33% of multiple tumours or 15% of bilateral neoplasms may be associated with concurrent tuberous sclerosis [2,3].

Upon microscopy, a triphasic, classic neoplasm constituted of myoid, spindle shaped cells, mature adipose tissue and dysmorphic, thick walled vascular articulations devoid of elastic lamina is observed.

Smooth muscle component is engendered from walls of blood vessels and may appear as hyper cellular, atypical, pleomorphic or demonstrate an epithelioid morphology.

Vascular component is configured of thick walled, hyalinised vascular articulations.

Adipose tissue component is constituted of aggregates of mature adipose tissue and may be encountered in a majority (> 90%) of tumours [2,3].

Multinucleated cells and multi-lobed nuclei are frequently observed. Focal haemorrhage, mitotic figures and zonal necrosis are frequently enunciated [3,4].

Epithelioid variant of angiomyolipoma may occur as a 'pure' form or may be composed of predominant population of polygonal cells pervaded with clear or densely eosinophilic cytoplasm and enlarged, hyperchromatic, bizarre nuclei.

Angiomyolipoma with epithelial cysts (AMLEC) represents as a morphological spectrum of neoplasms which characteristically delineate cysts layered with cuboidal or hobnail epithelial cells, simulating renal tubular epithelial cells.

Upon ultrastructural examination, an amalgamation of premelanosomes may be encountered [3,4]

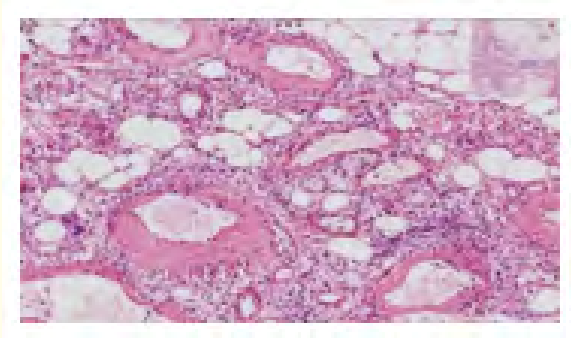

Figure 1: Angiomyolipoma composed of smooth muscle fragments intermingled with aggregates of mature adipose tissue and numerous vascular articulations. Mitotic activity and zonal necrosis is absent [6].

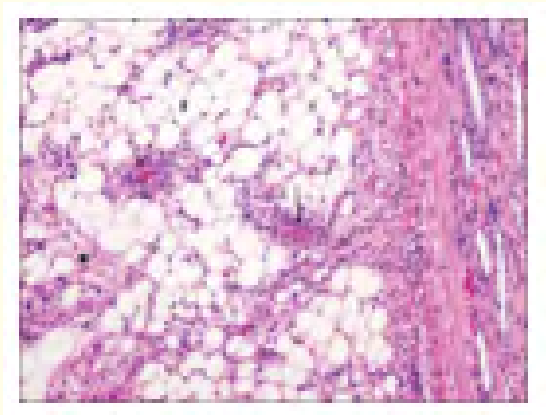

Figure 2: Angiomyolipoma exhibiting aggregates of mature adipose tissue cells commingled with fragments of smooth muscle and numerous patent vascular articulations. Intervening fibro-connective tissue septa are infiltrated by patchy chronic inflammatory cells [7].

Co expression of melanocytic markers confined to myoid component and smooth muscle immune markers confined to lipoid component is observed.

Angiomyolipoma appears immune reactive to human melanoma black 45 (HMB45) antigen, MART1/MelanA, cathepsin K, smooth muscle actin (SMA), muscle specific actin (MSA), calponin and caldesmon.

Angiomyolipoma appears immune non reactive to cytokeratin, PAX8, CAIX, GATA3, inhibin and CD117 [4,5].

Angiomyolipoma requires segregation from neoplasms such as clear cell variant of renal cell carcinoma, well differentiated liposarcoma, leiomyoma, leiomyosarcoma, pleomorphic rhabdomyosarcoma, malignant melanoma, adrenal cortical carcinoma, oncocytoma or mixed epithelial and stromal tumour of the kidney (MEST) [4,5].

Classic angiomyolipoma can be suitably discerned with cogent imaging. Besides, precise microscopic evaluation may adequately discern the neoplasm.

Appropriate classification of 'fat poor' and epithelioid variant of angiomyolipoma may be achieved with pertinent immunohistochemistry.

Cogent biochemical or haematological parameters appear within normal limits.

Upon imaging, characteristic radiographic appearance of classic angiomyolipoma delineates a prominent component of mature adipose tissue. Radiographic features are contingent to quantifiable mature adipose tissue constituting the neoplasm [4,5].

Angiomyolipoma may be appropriately treated with embolization or surgical extermination of the neoplasm.

Adoption of m TOR inhibitors as everolimus appear satisfactory for treating enlarged neoplasms extending into inferior vena cava.

Classic variant of angiomyolipoma pursues a benign clinical course. Tumefaction depicting epithelioid cellular component or pleomorphic morphological features are accompanied by an aggressive biological course [4,5].

Tumours delineating sarcomatous metamorphosis or distant metastasis are extremely exceptional. Retroperitoneal haemorrhage is encountered within a significant proportion of neoplasms. Bilateral neoplasms are associated with renal failure.

Tumour extension into contiguous organs, especially vascular articulations is associated with tumour associated mortality [4,5].

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6. Image 1 Courtesy: You tube.

7. Image 2 Courtesy: Journal of clinical imaging science.