Acta Scientific PAEDIATRICS (ISSN: 2581-883X)

Volume 6 Issue 6 June 2023

Case Review

Complaisant and Vascular-Littoral Cell Angioma

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Received: April 12, 2023

Published: April 25, 2023

Littoral cell angioma of spleen is an exceptionally discerned, benign, vascular neoplasm. Tumefaction characteristically originates from littoral cells which layer red pulp sinuses of spleen.

Littoral cell angioma displays significant concurrence with diverse immunological and neoplastic disorders. Clinical representation varies from asymptomatic tumours to incidental discovery or neoplasms with cogent clinical symptoms as pyrexia, pain and hypersplenism related cytopenias.

Neoplastic cells manifest with dual cellular differentiation of endothelial cells and histiocytic cells, reminiscent of normal splenic littoral cells. Characteristically, tumour cells appear immune non reactive to CD34 whereas diverse endothelial cell antigens may be expressed.

Littoral cell angioma commonly incriminates adults or middle aged subjects. Median age of disease emergence is 50 years although the neoplasm may appear within infancy to 9th decade. An equivalent gender predilection is observed [1,2].

Categorically, littoral cell angioma is confined to the spleen.

Of obscure aetiology, immune dysregulation is posited to expound clinical association of littoral cell angioma with diverse malignant neoplasms and immune mediated or autoimmune disorders.

Littoral cell angioma of spleen is a benign tumefaction wherein malignant metamorphosis is extremely exceptional. Around 50% lesions are asymptomatic [1,2].

Nearly 50% of incriminated subjects represent with abdominal pain, pyrexia, loss of weight, splenomegaly and hypersplenism associated thrombocytopenia and anaemia. Besides, neoplasm manifests with immune, autoimmune or metabolic disorders as Crohn's disease or immune thrombocytopenic purpura. Up to 60% tumours are associated with malignant disorders as colonic adenocarcinoma, pancreatic neoplasms, pulmonary carcinoma, renal cell carcinoma, lymphoma, myelodysplasia, aplastic anaemia or seminoma testis [2,3].

Upon gross examination, a solitary or multiple, distinctive nodules appear confined to splenic parenchyma. Multiple to solitary tumour nodules manifest a proportion of ~1:0.3 [2,3].

Neoplastic lesions emerge as miniature to enlarged nodules of variable consistency and hues ranging from yellow, brown, reddish or black, contingent to quantifiable necrosis, thrombosis, fibrosis and configuration of cysts. Alternatively, tumefaction may be spongy, cystic and well demarcated although devoid of a fibrous capsule [2,3].

Upon cytological examination, three dimensional clusters and cellular aggregates of bland, epithelioid or foamy cells demonstrating minimal nucleocytoplasmic ratio are discerned. Tumour cells may be pervaded with intracytoplasmic hemosiderin pigment [2,3].

Upon microscopy, neoplasm expounds proliferation of tortuous, anastomosing vascular articulations permeated with red blood cells. Vascular channels depict irregular lumens with frequently occurring papillary projections and cystic spaces. The vascular articulations are layered by tall endothelial cells which exhibit variable hemophagocytosis. Endothelial cells may slough into vascular spaces. Foci of sclerosis or cytological atypia are absent [3,4].

Littoral cell angioma is immune reactive to dual markers as endothelial cell markers factor VIII, CD31, von Willebrand factor and histiocytic markers as CD4, CD68, CD163, FLI1, MAC387, HAM56 and lysozyme. Besides, tumefaction is immune reactive to langerin (CD207), CD21, vimentin, cyclin D1 and variably immune reactive to S100 protein.

Ki67 proliferation index is minimal [4,5].

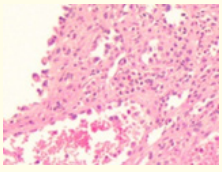

Figure 1: Littoral cell angioma depicting vascular channels lined with tall endothelium configuring papillary projections, cystic spaces, foci of hemophagocytosis and irregular lumens [6].

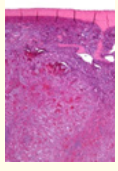

Figure 2: Littoral cell angioma delineating vascular articulations lined by tall endothelium, irregular lumens, papillary projections, cystic spaces and foci of hemophagocytosis [7].

Littoral cell angioma is immune non reactive to CD34, ERG, WT1, CD45, CD8, CD117, human herpes virus 8 (HHV8), CD1a and cytokeratin.

Littoral cell angioma is devoid of specific, diagnostic molecular features.

Littoral cell angioma of spleen requires distinction from neoplasms such as angiosarcoma, hamartoma, Kaposi sarcoma, splenic littoral cell hemangioendothelioma, haemangioma, hemangiopericytoma or lymphangioma [4,5].

Splenic littoral cell angioma manifests with splenomegaly along with tumour nodules constituted of benign, tortuous vascular channels. A dual composition of endothelial cells and histiocytic cells immune non reactive to CD34 is exemplified by the neoplasm.

Haematological assessment exhibits thrombocytopenia and anaemia in a subset of incriminated subjects [4,5].

Upon radiography, an enlarged spleen is encountered. Ultrasonography demonstrates a variable representation with heterogeneous splenic echotexture in the absence of specific nodules or configuration of hyper-echogenic, hypo-echogenic or iso-echogenic lesions.

Computerized tomography (CT) exemplifies solitary or multiple tumour nodules with hypo-attenuation [4,5].

T1 and T2 weighted magnetic resonance imaging (MRI) exhibits hypodense lesions on account of hemosiderin pigment deposition.

Littoral cell angioma is appropriately treated with splenectomy [4,5]. Subsequently, potential complications of asplenia as severe infection or thrombosis require precise management. Additionally, clinical evaluation of coexistent neoplastic lesions or immune disorders is mandated.

Typically, the benign littoral cell angioma is associated with excellent prognostic outcomes following splenectomy [4,5].

Possible malignant metamorphosis may ensue, especially with massive splenomegaly within spleen weighing > 1,500 grams or > 20 centimetre magnitude along the long axis. Exceptional instances of tumour reoccurrence or metastasis within an accessory spleen are documented [4,5].

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6. Image 1 Courtesy: Science direct.

7. Image 2 Courtesy: Wikipedia