Editorial

Burl and bud- sclerosing angiomatoid nodular transformation

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Editorial

Sclerosing angiomatoid nodular transformation is a contemporary, exceptional, non-neoplastic, vascular lesion of undefined etiology arising from the spleen. Characteristically, neoplasm is comprised of confluence of multiple angiomatoid nodules encompassed within concentric distribution of collagen fibres. Initially described by Martel in 2004, the uncommon splenic lesion was previously designated as multinodular hemangioma or may be denominated as splenic hamartoma or cord capillary hemangioma. Singularly confined to the spleen, the benign tumefaction depicts a typical morphology of multiple angiomatoid nodules entangled within a fibrosclerotic stroma. The lesion can be appropriately alleviated with surgical intervention. Tumour reoccurrence remains undocumented.

Sclerosing angiomatoid nodular transformation of the spleen exhibits a female predilection with female to male proportion of 2:1. The neoplasm preponderantly emerges in adults between 30 years to 60 years with mean age of disease occurrence at 48 years.[1,2]

As sclerosing angiomatoid nodular transformation is singularly confined to splenic parenchyma, extra-splenic incrimination is absent.

Of obscure etiology and pathogenesis, splenic sclerosing angiomatoid nodular transformation is posited to arise from an exaggerated sclerotic and neo-angiogenic splenic reaction secondary to various pre-existing conditions as vascular lesions with consequent thrombosis, vascular infarcts or hamartoma.[1,2]

Majority of lesions appear asymptomatic and are discovered incidentally upon imaging or surgical intervention for diverse unrelated diseases. Symptomatic tumefaction is accompanied by nonspecific symptoms as abdominal pain, abdominal discomfort, abdominal fullness, nausea, vomiting, unexplained loss of weight, splenomegaly and pancytopenia. Upon gross examination, a singular, un-encapsulated, well circumscribed, solid, significantly multinodular tumefaction appears variably traversed with fibrotic bands.[1,2]

Typically, the un-encapsulated, well circumscribed tumefaction is composed of reddish-brown nodules with alternating, band-like stromal component. A centric, fibrous, stellate scar is frequent.[2,3]

Upon microscopy, multiple, well circumscribed or confluent angiomatoid nodules are disseminated within splenic parenchyma. The nodules appear encompassed within variably quantifiable fibrotic and sclerotic stroma. Occasionally, a perimeter of fibrinoid tissue may simulate a granuloma-like appearance. Tumour nodules represent an admixture of slit-like, spherical or irregular vascular spaces layered with bland, plump endothelial cells. Foci of cellular and nuclear atypia or necrosis are absent. Mitotic figures appear insignificant.[2,3] Innumerable, nodular erythrocytes appear admixed within the stromal component comprised of collagenous bands imbued with myofibroblasts, hemosiderin laden macrophages, mature lymphocytes and plasma cells. Nodules of splenic sclerosing angiomatoid nodular transformation are configured of variable vascular articulations which can be appropriately emphasized with immunohistochemistry. The vascular configurations recapitulate normal composition of splenic red pulp as capillaries, sinusoids and miniature venules.[2,3] Intervening stromal component appears fibrous to fibro myxoid and is constituted of bland fibroblasts and myofibroblasts. The stroma is imbued with an abundance of inflammatory cells as small lymphocytes, hemosiderin laden macrophages and polytypic plasma cells. Innumerable plasma cells immune reactive to IgG4 may demonstrate an elevated IgG4: IgG ratio. Nevertheless, serum IgG4 values appear within normal limits. Adjoining splenic parenchyma appears unremarkable. Ultrastructural examination depicts miniature vascular spaces layered by endothelial cells permeated with pinocytotic vesicles. Weibel-Palade bodies are absent.[2,3] (Figure 1, 2)

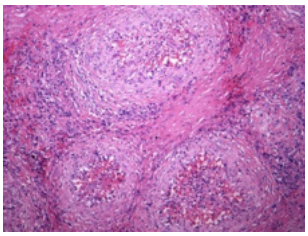

Figure 1 Sclerosing angiomatoid nodular transformation demonstrating angiomatoid nodules within splenic parenchyma composed of vascular articulations admixed with concentric collagenous stroma with infiltrating inflammatory cells as small lymphocytes, polytypic plasma cells, hemosiderin laden macrophages and myofibroblasts.

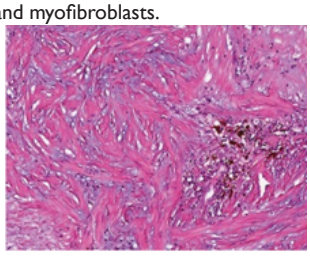

Figure 2 Sclerosing angiomatoid nodular transformation exhibiting vascularized parenchymal nodules intermingled with stromal inflammatory exudate of lymphocytes, plasma cells, hemosiderin laden macrophages and numerous myofibroblasts.

Sclerosing angiomatoid nodular transformation is configured of distinctive vascular articulations, as encountered within normal splenic red pulp. Capillaries configure an organized lobular pattern with immune reactivity to CD34+/CD8-/CD31+. Vascular splenic sinusoids exhibit an immune reactive pattern as CD34-/CD8+/CD31+ whereas miniature venules configure an intricate, mesh like pattern with CD34-/CD8-/CD31+ immune reactivity. Myofibroblasts confined to tumour nodules appear immune reactive to smooth muscle actin (SMA). Histiocytes or macrophages appear immune reactive to CD68. Constituent plasma cells manifest a polytypic expression of kappa and lambda light chains. Sclerosing angiomatoid nodular transformation of spleen appears immune non-reactive to D2-40. In situ hybridization may be beneficially adopted to discern Epstein Barr virus (EBV) infection.[3,4] Sclerosing angiomatoid nodular transformation requires segregation from neoplasms such as hemangioma, littoral cell angioma of spleen, hemangioendothelioma, angiosarcoma or hamartoma. Sclerosing angiomatoid nodular transformation of spleen simulates clinical and radiographic parameters of diverse splenic lesions, especially hemangioma, hamartoma or exceptional malignant neoplasms.[3,4] Commonly, hypervascular splenic lesions, congenital anomalies as accessory spleen, wandering spleen, asplenia, polysplenia, splenogonadal fusion, retrorenal spleen or benign mass lesions as splenic cyst, splenic pseudocyst, splenic haemangioma, splenic lymphangioma, splenic hamartoma, extramedullary hematopoiesis in the spleen, splenic abscess, splenic hydatid cyst, splenic inflammatory pseudotumor, splenic lipoma, splenic angiomyolipoma, splenic fibroma, sarcoidosis, focal splenic lesions in type I Gaucher disease, splenic hematoma or indeterminate mass lesions as solitary fibrous tumour, littoral cell angioma of spleen, inflammatory myofibroblastic tumour of spleen or malignant mass lesions as splenic lymphoma, angiosarcoma of spleen and splenic metastases from malignant melanoma or splenic malignant fibrous histocytoma, diffuse infiltrative processes manifesting with splenomegaly or distinct lesions as granulomas associated with splenic tuberculosis, splenic histoplasmosis, splenic siderosis, splenic amyloidosis or multifocal splenic lesions as lymphoma, distant metastases, sarcoidosis, fungal abscesses, granulomatous infections, splenic siderosis with Gamna-Gandy bodies or anomalies as traumatic or non-traumatic splenic rupture, splenic infarction, splenosis and splenic peliosis require exclusion.[3,4] Histopathological assessment of surgical resection specimen of spleen appears confirmatory. Core needle tissue sampling appears as a sensitive and specific technique of diagnosing haematological and non haematological lesions confined to spleen. However, complications as intraperitoneal seeding of undiscerned malignant neoplasms as angiosarcoma may be encountered.[3,4] Biochemical and hematological parameters appear unaltered. Occasionally, nonspecific features as elevated erythrocyte sedimentation rate, leucocytosis and polyclonal hypergammaglobulinemia may be encountered[4,5]

Neoplasm is devoid of characteristic or pathognomonic radiographic features. Ultrasonography, computerized tomography (CT) and magnetic resonance imaging (MRI) exhibits an isolated, solid, well circumscribed, multinodular tumefaction of variable magnitude with smooth or lobular perimeter. Ultrasonography characteristically depicts a well circumscribed, hypo-echogenic tumefaction.[4,5] Non contrast computerized tomography exhibits a homogeneous, iso-dense to mildly hypodense tumour nodule, in contrast to circumscribing splenic parenchyma. Post contrast computerized tomography demonstrates a hypo-vascular centric zone surrounded by an enhancing perimeter and radiating spokes of vascularized tissue extending from periphery towards centric lesion. Progressive centric image enhancement with delayed imaging may ensue on account of contrast medium penetrating centric lesion from vascularized rim, a configuration denominated as a 'spoke wheel' pattern.[4,5] T1 weighted magnetic resonance imaging exhibits a heterogeneous signal of mild to intermediate intensity. Typically, T2 weighted magnetic resonance imaging delineates minimal signal intensity, in contrast to diverse splenic lesions. T1 weighted imaging with gadolinium contrast enunciates peripheral and septal image enhancement with 'spoke wheel' pattern configuring a centric, hypo-enhancing stellate scar.[4,5] Fluorodeoxyglucose positron emission tomography(FDG/PET) appears hypermetabolic and demonstrates enhanced standard uptake values , in contrast to hepatic and circumscribing normal splenic tissue. Abundant cellular component as hemosiderin laden macrophages, myofibroblasts, lymphocytes and plasma cells may account for avidity upon FDG/PET scan. Upon scintigraphy, sclerosing angiomatoid nodular transformation appears cold. Absence of constituent reticuloendothelial cells may account for lack of uptake of 99mTc-sulfur colloid.[4,5] The condition can be appropriately alleviated with splenectomy which an optimal and recommended mode of therapy, as the neoplasm is preponderantly discovered incidentally.[4,5] Sclerosing angiomatoid nodular transformation is associated with superior prognostic outcomes. Following splenectomy, tumefaction appears devoid of localized reoccurrence. Malignant metamorphosis remains undocumented. [4,5]

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Conflicts of interest

The author declares that there are no conflicts of interest.

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