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Short Communication

The Misshaped Prevision- Splenic Hamartoma

Anubha Bajaj*

Department of Anatomy, Consultant Histopathologist, AB Diagnostics, India

***Corresponding Author:** Anubha Bajaj, Department of Anatomy, Consultant Histopathologist, AB Diagnostics, India.

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Splenic hamartoma is an extremely exceptional, benign neoplasm arising from splenic parenchyma. Splenic hamartoma is additionally designated as splenoma, splenoadenoma or nodular hyperplasia of spleen. Besides, the contemporary sclerosing angiomatoid nodular transformation (SANT) of spleen manifests as a non neoplastic, vascular lesion and may be contemplated as a fibrosing variant of splenic hamartoma. Commonly congenital, splenic hamartoma is constituted of an aberrant admixture of normal splenic tissue. The preponderantly nodular lesion of variable magnitude is derived from cells layering the splenic sinuses. Splenic hamartoma represents with clonal chromosomal anomalies. Splenic hamartoma demonstrates an equivalent gender predilection. No age of disease emergence is exempt [1,2]. Splenic hamartoma is discovered incidentally upon imaging or surgical intervention for diverse conditions or autopsy. Tumefaction may represent with pain or palpable nodule. Enlarged lesions may manifest with mass effect. Spontaneous rupture of spleen may ensue. Splenic hamartoma may be associated with tuberous sclerosis, Wiskott-Aldrich-like syndrome or hypersplenism demonstrating thrombocytopenia, anaemia or pancytopenia. Exceptionally, tumefaction may concur with malignant haematological conditions [1,2]. Upon gross examination, splenic hamartoma represents as a singular or multiple, nodular, circumscribed, dark red lesion which compresses encompassing splenic parenchyma. Tumour magnitude varies from few millimetres to several centimetres. Frequently, splenic hamartoma emerges as a solitary or multiple, spherical, well circumscribed, un-encapsulated, bulging tumour nodule which compresses abutting normal splenic parenchyma. Tumefaction may concur with an admixture of normal splenic architecture as white pulp or red pulp. Foci of fibrosis and cystic transformation may ensue [3,4]. Upon microscopy, splenic hamartoma may manifest a follicular tumour configuration. Alternatively, a pulpous or fibrous neoplasm may ensue. The well circumscribed tumefaction of variable magnitude is comprised of disorganized vascular articulations which appear commingled with splenic red pulp. Neoplasm appears to abut aggregates of entrapped mature adipocytes. Foci of extramedullary haematopoiesis may ensue [3,4].

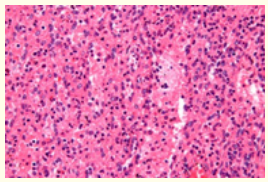

Figure 1: Splenic hamartoma delineating disorganized vascular channels lined with endothelial cells and perfused with red blood cells along with an admixture of fibrous tissue and mature adipose tissue [7].

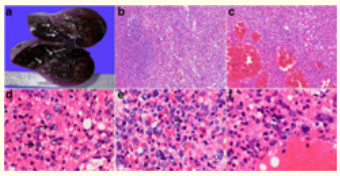

Figure 2: Splenic hamartoma exhibiting a dark red, spongy spleen constituted of disorganized vascular channels lined by plump endothelium with focal haemorrhage and an encompassing fibrotic stroma [8].

Splenic hamartoma is immune reactive to CD8, CD68 or smooth muscle actin (SMA). Reticulin stain may be employed to highlight the fibrous component. Endothelial cells may be stained with nonspecific esterases with **α naphthyl acetate** and naphthol acid phosphate as substrate. However, the cells are devoid of alkaline phosphatase. Immune reactivity to CD34 is variable [3,4]. Splenic hamartoma requires segregation from neoplasms such as angiosarcoma, haemangioendothelioma, splenic haemangioma, littoral cell angioma, splenic lymphangioma, inflammatory myofibroblastic tumour or sclerosing adenomatoid nodular transformation. Microscopic distinction from lesions such as haemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, sclerosing angiomatoid nodular transformation or angiosarcoma can be challenging and is necessitated. Nevertheless, splenic hamartoma is immune reactive to CD68, in contrast to diverse vascular lesions of spleen [3,4]. Upon imaging, splenic hamartoma demonstrates a focal, disorganized, excessive growth of splenic parenchyma with an echogenicity, attenuation and signal intensity akin to surrounding normal splenic parenchyma [5,6]. Upon ultrasonography, majority of splenic hamartomas appear as a hypoechoic, solid tumefaction. Nevertheless, a heterogeneous tumour mass may be discerned due to haemorrhage or cystic alterations. Upon colour Doppler and following contrast administration, the lesion appears hyper vascular [5,6]. Computerized tomography (CT) delineates splenic hamartoma as an is odense or hypodense, solid tumefaction which exemplifies heterogeneous image enhancement upon administration of contrast medium, as compared to adjacent normal splenic parenchyma. Magnetic resonance imaging (MRI) is an optimal, recommended imaging modality which appropriately differentiates splenic hamartoma from splenic haemangioma [5,6]. Upon T1 weighted magnetic resonance imaging, majority of lesions appear isointense [5,6]. Upon T2 weighted magnetic resonance imaging, majority of lesions appear heterogeneously hyper-intense [5,6]. Upon administration of gadolinium contrast, splenic hamartoma characteristically depicts an immediate, vivid, post contrast image enhancement, as compared to haemangioma [5,6]. Delayed post contrast imaging delineates a hamartoma with uniform and intense image enhancement along with or devoid of central hypo-vascular zones [5,6]. Splenic hamartoma can be appropriately treated with comprehensive or partial splenectomy. Additionally, minimally invasive techniques as laparoscopic or hand assisted laparoscopic modalities may be beneficially employed [5,6].

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7. Image 1 Courtesy: Wikimedia commons.

8. Image 2 Courtesy: Diagnostic pathology-biomed central.