(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 6, Issue 2 , Page No: 567-571
March-April 2023

IJMSCR

Splenic Nodular Hamartoma – A Rare Case Report With Review Of Literature

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Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

Splenic nodular hamartoma (SNH) is a rare benign tumor. It is usually congenital and present as a nodular lesion of variable size, derived from splenic sinus-lining cells and composed of aberrant mixture of normal splenic tissue. More than 150 cases of SNH have been documented in the literature till date. SNH is also called splenoma, splenoadenoma or nodular hyperplasia of the spleen. We discuss a case of SNH in a young female presenting with splenic mass lesion.

Keywords: SNH, SPLENIC NODULAR HAMARTOMA, SPLENOMA, SPLENIC HAMARTOMA

Introduction

SNH is a rare tumor with benign vascular proliferation of splenic sinus-lining cells and composed of aberrant mixture of normal splenic tissue. SNH is very rare, in a 17 years long study at Medical Centre only 3 cases were described in 200000 splenectomies [1] and an incidence of 0.024% to 0.13% was noted in a review of autopsies [2]. It affects all age groups with equal sex preponderance. It is usually an incidental finding but can present with pain, palpable mass or spontaneous rupture or may be associated with tuberous sclerosis, Wiskott-Aldrich like syndrome and hypersplenism, this condition is usually treated by complete and partial splenectomy [3]. We describe a rare case of splenic nodular hamartoma in a young female presenting with features of hypersplenism and splenic mass lesion.

Case report

A 35 year old woman G1P1L1 presented with pain in the left hypochondrium and generalized weakness, breathlessness and leg pain since two years. The upper abdominal pain is insidious in onset, progressive, dull aching in intensity with history of fever on and off. She also had history of blood transfusion four times in last two years. She doesn't have any past history of tuberculosis, diabetes and hypertension. On general examination she was conscious and well oriented and has marked pallor and pitting edema in bilateral lower limbs. On per abdomen examination liver was just palpable and moderate splenomegaly, firm in consistency and reaching till umbilicus was palpable. Other systemic examination was normal.

On investigating the patient complete hemogram revealed- Hb-6.8 gm/dl, hematocrit 21.3%, MCV-77.2fl, total leukocyte count was 3500/cumm and platelet count was1.5 lacs/ul. Abdominal ultrasonography was performed, showing normal liver, biliary ducts, and pancreas morphology. Nevertheless a 17.4 cm enlarged spleen was found, containing rounded well defined hyperechoic solid mass measuring 8x8.5 cm with diffuse calcification and on Color Doppler showed increased blood flow. On clinicoradiological correlation clinical diagnosis

of hypersplenism with splenic mass was made and she underwent splenectomy.

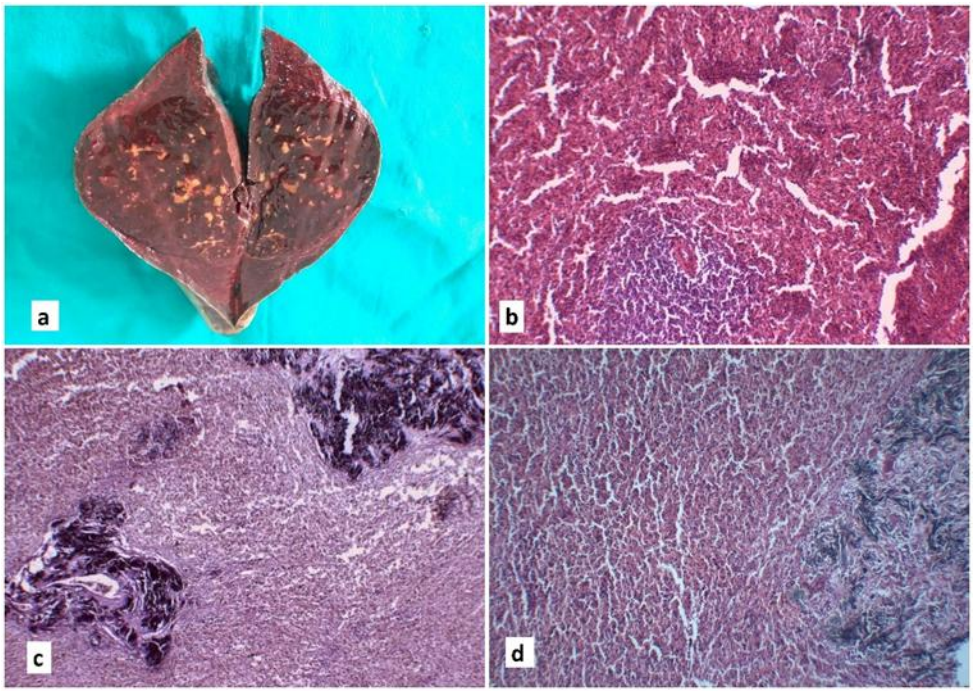
Gross examination revealed splenectomy specimen measured 15x 14x10 cm and weighing 862 gm and serial sectioning showed presence of single thinly encapsulated fleshy reddish brown nodular lesion, compressing splenic parenchyma measuring 8.5x8x7 cm (Figure 1a). On microscopy, H&E stained sections examined from lesion show mixture of disorganized blood vessels of varying sizes intermingled with splenic red pulp element and areas of fibrosis with numerous Gamna Gandy bodies (Figure 1b-d). The rest of the spleen show both red pulp and white pulp with mild congestion. The histomorphological features were consistent with splenic nodular hamartoma.

Figure 1a- Photograph of the cut surface of the resected spleen illustrating well-circumscribed bulging solid mass compressing the rest of the spleen

Figure 1b- H&E stained section show a non- encapsulated hamartoma on the right and normal splenic parenchyma on the left (100X)

Figure 1c- H&E stained section show mixture of disorganized blood vessels intermingled with areas of fibrosis and show numerous gamma gandy bodies. (40X)

Figure 1d-High power view of the lesion show vascular slits lined by occasionally plump endothelial cells (100X)



Discussion

Solid lesions of the spleen are very rare and generally asymptomatic. About half of these are accidentally detected during imaging studies carried out for unrelated causes [4]. Differential diagnosis includes primary malignancies, in particular non-Hodgkin's lymphoma and angiosarcoma, which is the most common non lymphoid malignant primary tumor of the spleen [5]. Lung cancer, melanoma, ovary, uterine cervix, and other non-gastrointestinal tumors showing splenic metastasis were also reported [6, 7]. Benign lesions are extremely rare (7/100000 autopsies) and have generally of vascular origin [8]. Hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, and hamartoma of the spleen have been previously described in the literature [8-14]. Since the first report in 1861 by Rokitansky [15], splenic hamartomas have also been called splenomas, spleen within a spleen, hemangiomas, posttraumatic scars, fibrotic nodules, tumorlike congenital malformations, and hyperplastic nodules [16-18].

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Splenic hamartomas are very rare. Now a day's improvement in imaging techniques have led to increased detection of this entity and a secondary rise in incidence [15]. Although hamartomas are benign and usually asymptomatic, it is important to distinguish this benign lesion from malignancy.

Clinical Features- Splenic hamartomas can occur in any age group (11 months to 86 years) [17] with equal occurrence in males and females, usually without symptoms[18]. Its size tends to be longer in females, probably due to hormonal influence on tumor growth [19-21]. The tumor is usually detected as a singular lesion with a diameter ranging from a few millimeters to several centimeters. Symptoms such as pain, palpable mass or spontaneous rupture are associated with longer lesions. Hypersplenism, including thrombocytopenia, anemia, pancytopenia, or malignant hematologic conditions, is described even if uncommon [22-24]. Most splenic hamartomas are hyperechoic solid masses, with or without cystic changes in ultrasonogram, and are hypervascular in both color Doppler ultrasound and angiogram. On computed tomography, hamartomas appear as isodense or hypodense solid masses and demonstrate heterogeneous contrast enhancement relative to adjacent normal parenchyma [25]. Most of the lesions are isointense in T1-weighed MRI and heterogeneously hyperintense in T2-weighed MRI [25, 26]. Although splenic hamartoma may be suggested by radiologic findings, definitive diagnosis requires tissue examination. Splenectomy is curative for rare symptomatic hamartomas, but a partial splenectomy may be sufficient in some cases [27].

Hamartomas are solitary or multiple, round, well-circumscribed, unencapsulated bulging nodules compressing the adjacent normal splenic parenchyma. Focal fibrosis and cystic areas can be seen. The color is usually dark red to grayish white. The size ranges from a few millimeters to centimeters, with a median size of 5 cm, but lesions as large as 20 cm have been reported [17,18]. Histologic findings reveal disorganized vascular channels lined by slightly plump endothelial cells without atypia, mixed with intervening splenic red pulp–like stroma with or without white pulp [19, 20]. Gamna-Gandi bodies were recognized in the splenic parenchyma removed from the nodule were very uncommon. Some large splenic hamartomas have been associated with infarcts and siderotic bodies [2, 11, 28].The latter can be explained solely on the basis of large spleen size and are probably not related to the hamartoma.

In our case, a young female patient was presented with clinical features of hypersplenism and radiologically detected rounded well defined hyperechoic vascular solid mass and she had underwent splenectomy and gross splenectomy specimen showed single nodular bulging lesion with characteristic histological findings and numerous Gamna-Gandi bodies.

The pathogenesis of hamartoma is controversial. Some consider hamartomas as congenital malformation of the splenic red pulp, a neoplasm, excessive and disorganized growth of abnormally formed red pulp, or a reactive lesion to prior trauma [2, 7]. With documented cases associated with hematologic malignancy, others believe hamartoma is an acquired proliferative process [11]. Splenic hamartomas must be differentiated from other vascular tumors of the spleen, including hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, sclerosing angiomatoid nodular transformation of the spleen and angiosarcoma. Solid mass–forming lesions of the spleen, such as inflammatory myofibroblastic tumor, lymphoma, metastatic disease, disseminated fungal or mycobacterial infections, and sarcoidosis are also included in the radiologic differential diagnosis [29].

Splenic hemangiomas are the most common benign neoplasm arising from sinusoidal epithelial cells. The cavernous type is more common than the capillary type. Histologically, hemangioma is composed of proliferating vascular channels, which are lined by flat endothelial cells and separated by thin fibrous septa or red pulps [25]. The flat endothelial cells are positive for endothelial markers including CD31 and CD34 and are negative for CD8, CD21, and CD68.

Littoral cell angioma is a vascular tumor arising from littoral cells originating from splenic sinuses. Littoral cells are characterized by their expression of both endothelial and histiocytic markers. Histologic examination demonstrates anastomosing vascular channels lined by tall columnar cells, which often show hemophagocytosis. The lining cells are positive for both endothelial and histiocytic markers, CD31 and CD68 [25, 26]. CD21, which is the C3d complement receptor and also the Epstein-Barr virus

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receptor of B lymphocytes, is reported to be exclusively positive in littoral cell angioma. Unlike splenic hamartoma, littoral cell angioma is negative for CD8 and CD34 in the lining cells [25, 30].

Sclerosing angiomatoid nodular transformation of the spleen, also known as multinodular hemangioma, is altered red pulp entrapped by non-neoplastic stromal proliferation [27]. Microscopically, the lesion is composed of multiple confluent vascular nodules surrounded by concentric collagen fibers or fibrinoid rims. The central portion of the nodules consists of vascular channels of varying caliber lined by plump endothelial cells interspersed with ovoid or spindle cells. Immunostaining of the vascular area in spleen reveals 3 types of blood vessels: CD34-CD31+CD8+ in sinusoids, CD34+CD31+CD8- in capillaries, and CD34- CD31+CD8-small veins. Scattered macrophages are positive for CD68.

Despite its similarity with splenic hamartoma in composition, sclerosing angiomatoid nodular transformation is a mixture of 3 types of blood vessels, whereas hamartoma consists of only sinusoid-type vessels.A key immunohistochemical feature is CD8 positivity of the lining cells of the vascular channels. The cells are also positive for CD31, factor VIII–related antigen, and vimentin [2, 9, 12, 19]. Immunostaining results for the lining cells with CD34 have been inconsistent [2, 18, 20, 22].

Conclusion:

With the rapid advancement of imaging modalities, smaller asymptomatic lesions of the spleen are being identified. Splenic hamartoma is a benign vascular proliferation characterized by CD8 positive immunophenotype of the lining endothelial cells. We should be aware of this rare, benign entity and interpret histologic features with clinical and radiologic findings to give a diagnosis and differentiate it from malignant lesions of spleen.

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