A CASE OF SPLENIC HAMARTOMA ASSOCIATED WITH THROMBOCYTOPENIA

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Abstract

Splenic hamartoma is very rare and benign tumor of the splenic. The few cases have been reported in the literature to date, this usually discovered incidentally during a radiological examination or an operation. We report a case of 49-year-old female with no past medical history, presented 8 month ago a pain and discomfort abdominal related to the growth mass in the left hypochondrium. Physical examination was remarkable a firm and painless mass below the left costal margin measuring almost 15 cm. Laboratory tests revealed a platelet count to 95000/mm. The computed tomography showed heterogeneous splenomegaly (19x14 cm) enhancement of the mass in the arterial phase and containing multiple areas of necrosis. A splenectomy was performed. Histologic study concluded to the diagnosis of splenic hamartoma.

Introduction:-

Splenic hamartoma are rare, non-neoplastic tumors (1). Only about 150 cases have been reported since its first description by Rokitansky in 1861 (2). Most patients are asymptomatic and splenic hamartomas are usually discovered incidentally on imaging and few have symptoms of hypersplenism as thrombocytopenia, anemia and pancytopenia. We are representing a case of splenic hamartoma associated with thrombocytopenia.

Case Report:

A 49-year-old-female patient was admitted to the hospital with a 8-month history of pain and discomfort abdominal associated to the progressive growth of volume of the left side, his medical story was without notable pathological; on examination, the patient was hemodynamically stable, afebrile and the normal conjunctives, physical examination revealed a firm and painless big mass below the left costal margin measuring 15 cm, without any other associated sign. Laboratory studies showed a hemoglobin of 13.2, a platelet count a 95 000 and white count a 7650. Abdominal computed tomography (TC) was performed, and images before the administration of contrast material showed an increased spleen size (19 cm), seat of a voluminous, rounded upper medio-polar mass well limited, hypodense, raised heterogeneously after injection and containing multiple areas of necrosis, this mass measures 15x11 mm and extended over 14 cm in height. This mass adjoins the left diaphragmatic dome and comes into contact with the stomach and the left hepatic lobe within, has close contact with the left adrenal and the upper pole of the left kidney, without evidence of infiltration. and the diagnosis of splenic hamartoma was tackled with these radiological characteristics.
Splenectomy was eventually performed in laparotomy with careful dissection. The patient evolved favorably and was discharged on her fourth postoperative day.

A pathological examination of the specimen revealed the 25x 14 cm spleen, weighing 900 g, the presence of well limited cystic cavity has coagulated hemorrhagic content without endocystic vegetation measuring 10x7x6 cm.

At microscopic examination of specimen of the spleen and immunohistochemistry was consistent with the diagnosis of splenic hamartoma. The patient was seen at the ambulatory clinic six months after her surgery with an auspicious evolution, presenting normal hemoglobin and platelet levels.

Figure 1: Abdominal computed tomography revealed a kid (14cm) hypodense splenic mass containing multiple areas of necrosis (a). In coronal cut voluminous splenomegaly and mass compress the left diaphragmatic dome, stomach and the left kidney (b).
Discussion:
The concept of hamartoma was formulated in 1994 by Albrecht (3) a non-neoplastic tumor-like congenital malformation consisting of a variable mixture of tissues normally present in the organ affected, sometimes with an overgrowth of one or more of them (4).

Hamartoma are rare benign tumors of the spleen, composed of an aberrant mixture of normal tissue components. Forming single or multiple masses within the splenic pulp (1). It can occur in any age group (5 months to 86 years) with a mean of age of 47 years (5) and with no sex predilection (6), woman tend to have larger lesions, probably due to hormonal factors (7) no statements can be proven. More than 80% of patients with splenic hamartomas are asymptomatic and the lesions are usually detected during imaging studies, surgery or post mortem (5,8), sometimes few cases have symptoms such as pain, palpable mass or splenomegaly may also cause anemia, thrombocytopenia or pancytopenia or malignant hematological conditions, has also been reported (9). Moreover, our patient presented with low thrombocytopenia level, due to the important of splenomegaly she presented, returning to her normal level after surgery was performed. other less common symptoms include fever, night sweats, malaise and spontaneous rupture (5). They may also be associated with multiple hamartomatosis, tuberous sclerosis and with Wiskott-Aldrich-like syndromes and Kasbach-Merrit syndrome (10,11).

Imaging of splenic hamartomas may be not specific. At abdominal ultrasound they usually appear as solid, homogeneous hypoechoic mass (10), with positive vascularization at color Doppler (12), sometimes with multiple anechoic cystic changes or with an inhomogeneous appearance (10). They can also appear as an isodense mass on CT. after contrast medium administration, these masses show early and sustained enhancement during the delayed phase of contrast-enhanced CT (8). Clinical or imaging diagnosis of this type of tumor is difficult to make before the operation, nevertheless, two documented cases succeed in having a preoperative diagnosis (13,14) us our patient. At MRI hamartomas are isointense on T1 weighted images and hyperintense on T2 weighted images, with heterogeneous enhancement after gadolinium administration (15,16).

The vascular tumors of the spleen represent the main differential diagnosis of this disease including hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, sclerosingangiomatoid nodular transformation of the spleen, and angiosarcoma. Solid 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 (7). Diagnosis is confirmed by histopathological examination.

There are three histologically types of splenic hamartoma: the pulposal type, resembling the splenic red pulp, the lymphoid type or follicular, resembling the splenic white pulp, and the fibrous type (17). Immunohistochemistry may reveal CD8 positive cells lining the vascular channels (18). These cells are also positive for CD31, factor VIII–related antigen, and vimentin. Immunostaining for CD34 has led to inconsistent results, and the endothelial cells are negative for CD21. CD68 is positive in scattered stromal macrophages but negative in the cells lining the vascular channels (7).

Currently, laparoscopy splenectomy is become a standard for treatment of most of splenic disease (19). But open splenectomy is considered better indicated in case of splenomegaly, when the diameter of spleen exceeds 20 cm (20). In the present case, the length of spleen was 19 cm, therefore we have adopted an open approach due to the presence of thrombocytopenia, the risk of spontaneous splenic rupture and its vascularization. The prognosis of splenic hamartomas is good, as local and distal recurrence rate is very low and splenectomy can therefore be considered curative (21).

Splenic hamartomas associated with thrombocytopenia have rarely been reported. Komo et al (22) report that only 19 cases have been reported in literature and only 6 cases have been reported in the Japanese literature. Watanabe et al (23) reported that the mean diameter of splenic hamartomas was 5.3 cm, and the diameter of those associated with thrombocytopenia was 9.9 cm. Furthermore, they reported that thrombocytopenia was caused by hypersplenism due to splenomegaly. In the present case, thrombocytopenia caused by hypersplenism due to splenomegaly of splenic hamartoma, the histological examination not showed any sign of myelodysplastic syndrome.

Conclusion:-
this observation reports the case of splenic hamartoma associated with hematological disorders in particular thrombocytopenia, the preoperative diagnosis can be suspected by scanner. Surgery can confirm the final diagnosis and resolves the symptoms related to hypersplenism.

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