

Hemangioma with Littoral Cell of the Spleen: About A Case

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ABSTRACT

Splenic hemangioma is a benign vascular lesion of the spleen that develops at the expense of red pulp structures. It is most often discovered accidentally, radiologically. It often raises a differential diagnosis problem with splenic hamartoma or splenoma, which is defined as a nodular formation consisting of the same elements of normal splenic parenchyma but arranged in a haphazard way. We report the case of a 50-year-old woman hospitalized for pain of the left hypochondrium.

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Introduction

Case report

50-years-old woman, single. She had a personal history of hysterectomy 5 years ago for poly-myomatous uterus. In her family history she had a brother operated on for a hydatid cyst of the liver.

The patient had intermittent pain in the left hypochondrium for 8 months with moderate intensity without specific irradiation and no accompanying gastrointestinal signs. Everything evolves in a context of conservation of the general state and apyrexia.

The physical examination found a patient in good general condition, hemodynamically stable, apyretic with normo stained conjunctiva. The abdominal examination found isolated splenomegaly of 14cm, slightly painful on palpation. There was no peripheral lymphadenopathy, nor collateral circulation, nor hepatomegaly.

The biological assessment revealed a correct hemoglobin level at 13.2g / dl, white blood cells at 7650 element / mm³ and thrombocytopenia at 95000 element / mm³. The C-reactive protein was 20. There were no hydroelectrolytic disorders, the renal and hepatic check was without abnormality. The tumor markers were normal.

The abdominal ultrasound showed an enlarged spleen, with regular contours, seat of a heterogeneous, hyperechoic mass containing cystic formations, measuring: 136 * 138 * 113mm, without signs of PH. IMAGE 1.

The CT scan revealed an enlarged spleen (19cm), with a large, rounded, well-defined, hypodense, medio-polar mass, heterogeneously enhanced after injection and containing multiple areas of necrosis. This mass measures 15 * 11mm and extended on 14cm of height. It remains at a distance from the splenic hilum, contiguous to the left diaphragmatic cupola and to the middle arches of the 7th to the 10th left rib, it comes into contact with the stomach and the left hepatic lobe inwards, without signs of infiltration. With respect for peripheral fat. Without visualization of ADP or associated

ganglion. She concluded with a splenic Hamartoma. IMAGE 2

Hence the indication of splenectomy by laparotomy. The splenectomy piece weighed 900g and measured 25 * 14 * 10cm. With the cut presence of a cystic cavity well limited coagulated haemorrhagic content, without endocystic vegetation measuring 10 * 7 * 6cm encapsulated pace. IMAGE 3

Anatomopathological study showed a tumor proliferation made of vascular section with sometimes cystic haematological content. These slits are lined by numerous regular endothelial cell sites devoid of atypia and mitotic figure. These cells are recognized by the CD31, CD34 and CD68 antibody in the immunohistochemical study. IMAGE 4 AND 5.

This morphological and immunohistochemical aspect favored a splenic littoral hemangioma of 10 cm long axis, without vascular or capsular invasion.

The follow-up was simple and no complication was reported.

Discussion

Splenic hemangioma with littoral cells is a rare primary vascular tumor of the spleen, described for the first time by Falk et al (1) in 1991. It is a benign tumor but rare cases of malignancy have been reported (littoral cell angiosarcoma) (2). It develops from littoral cells, which normally line the splenic sinuses of the red pulp of the spleen.

It occurs at any age with no predominance of sex. Clinically it can manifest as isolated splenomegaly, sometimes signs of hypersplenism can be found. But it is most often asymptomatic of chance discovery (3).

Biologically thrombocytopenia is almost constant.

On the ultrasound, the splenic hemangioma appears in the form of nodules that can be iso-echogenic, hypoechogenic or hyperechogenic within a spleen having a marbled appearance (4). In its mixed form (hypo and hyper echogenic) with pseudocystic elements will arise the differential

diagnosis with the hamartoma of the spleen (5). Doppler has a central and peripheral vasculature (4,5)

CT characterization of the littoral cell hemangioma reveals a hypodense nodule within a hypertrophied spleen, ranging from 5 to 6 cm in size and presenting contrast enhancement of the portal venous phase. At late time nodules become isodense to surrounding splenic parenchyma due to late filling of nodules. (3,6)

The radiological results can rarely lead to a definitive diagnosis because many differential diagnoses including splenic hamartoma have the same radiological aspects.

It is the anatomopathological and immunohistochemical study that makes the certainty diagnosis.

Macroscopically, the spleen has on its surface one or more nodules with blood of varying color ranging from dark red to brown through black, depending on the chronicity of the blood in the lesion.

Histologically, littoral cell angioma is characterized by a proliferation composed of vascular spaces of variable size, interconnected, with long papillary projections in their lumen.

They are lined with two types of cells. The former are tall endothelial cells and are in contact with the lights. They have a tendency to exfoliation in vascular lumens. The second, basal, are endotheliiform, located under the first in contact with the underlying chorion. The lesions do not have a capsule but are well delimited from the surrounding splenic parenchyma. (7)

The particularity of the splenic hemangioma with littoral cell is that it presents to the immunohistochemical study a double positivity at the same time to the vascular and macrophagic markers CD31 and CD68 / KP1. The basal component also expresses CD34 (8).

Conclusion

Hemangioma with littoral cell of the spleen is a rare benign splenic tumor composed of multiple vascular channels filled with blood. Isolated splenomegaly is the major sign. Angioma with littoral cells must be taken into account in the differential diagnosis of several splenic masses. His definitive diagnosis is made by anathomopathology and immunohistochemistry.



IMAGE1. Increased rate of size, regular contours, seat of a heterogeneous mass, hyperechoic containing cystic formations.

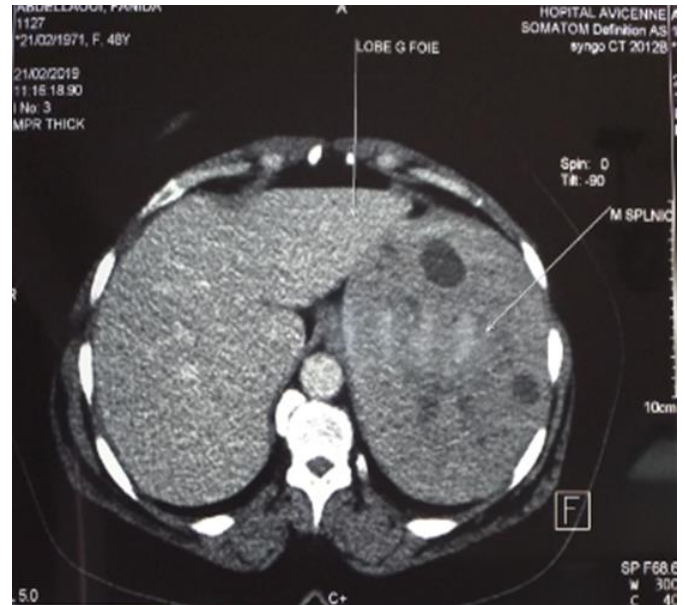
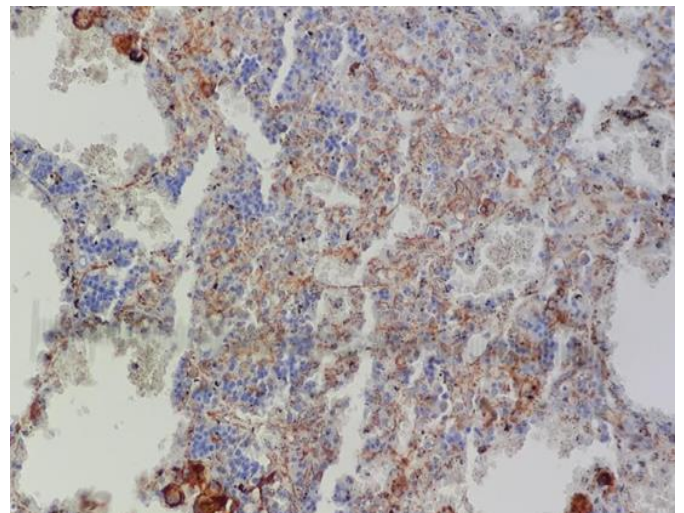


Image 2. Increased size (19 cm), seat of a large, rounded, well-defined, hypodense, medio-polar mass, heterogeneously enhanced after injection and containing multiple necrotic.



Image 3. The splenectomy piece weighed 900g and measured 25 * 14 * 10cm.



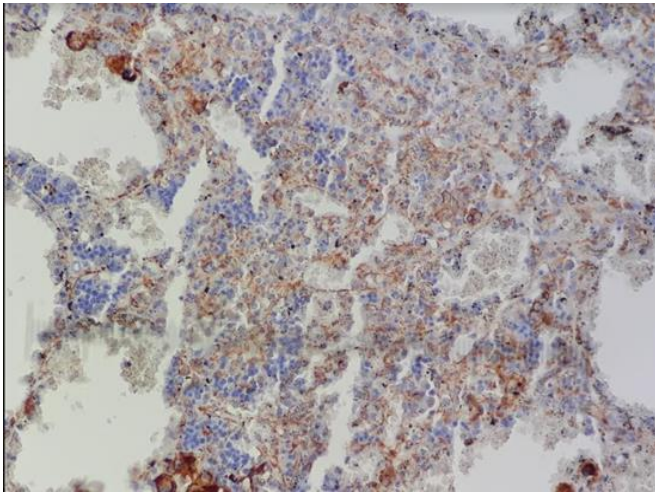


Image 4 and 5. Immunohistochemical profile of a CD31-positive CD34 negative and CD68 positive splenic littoral angioma in the stroma histiocytes.

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