

SPLENIC HEMANGIOENDOTHELIOMA IN PAEDIATRIC PATIENTS. CASE REPORT

Hemangioendotelioma esplénico en paciente pediátrico: Presentación de caso

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Palabras clave (DeCS)

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Summary

Splenic hemangioendothelioma are rare vascular neoplasms that occur in patients between the ages of 3 and 54. Few pediatric cases have been described. It is characterized by well-circumscribed lesions, properly separated from the healthy splenic parenchyma, moderate cellular atypia, little mitotic activity, and an intermediate histological appearance between hemangioma and angiosarcoma.

Resumen

Los hemangioendoteliomas esplénicos son neoplasias de origen vascular poco frecuentes, se presentan en pacientes entre 3 y 54 años. En edad pediátrica se han descrito pocos casos. Se caracteriza por presentar lesiones bien circunscritas, debidamente separadas del parénquima esplénico sano, moderada atipia celular, poca actividad mitótica y una apariencia histológica intermedia entre el hemangioma y el angiosarcoma.

Introduction

The term hemangioendothelioma (HE) has been used to describe a heterogeneous group of vascular neoplasms, which are intermediate between benign and malignant tumors (1).

Primary HE of the spleen is rare (1,2). It is a tumor of intermediate aggressiveness, between benign hemangioma and highly malignant HE. It is a mildly malignant vascular tumour and very rare (3,4).

To date, only a few cases of splenic HE have been reported in adults (1). Although its aetiology is unknown, the average age is close to 25 years, ranging from 3 to 54 years, with male predominance 4:2 (5). In paediatric patients, only two cases have been described in Latin America until 2014 (1).

Case report

A 4-year-old male patient from southern Honduras with a personal history of cutaneous leishmaniasis six months ago, presents to the paediatric

emergency service with a history of abdominal pain and distension, accompanied by asthenia and adinamia.

On physical examination she is underweight for her age, pain on palpation in the upper left quadrant, enlarged spleen two centimeters below the rib ridge.

Laboratory tests were carried out in which no alteration was evident. Ultrasound yielded the following results: spleen with regular contours, increased size (8.4 x 3.1cm), with mass of mixed characteristics of solid predominance, defined edges, heterogeneous. When examining with Doppler color presents central and peripheral flow. Its dimensions of 4 x 4,3 x 4,2 cm, with anechoic central area related to necrosis (figure 1). In the tomography (CT) the following findings were found: hypervascularized solid mass, located in the upper pole and middle third of the spleen, with irregular edges, with necrotic center, of 4,7 x 4,3 x 4,6 cm, in arterial phase presents prominent enhancement, with moderate washing in venous phase (figure 2).

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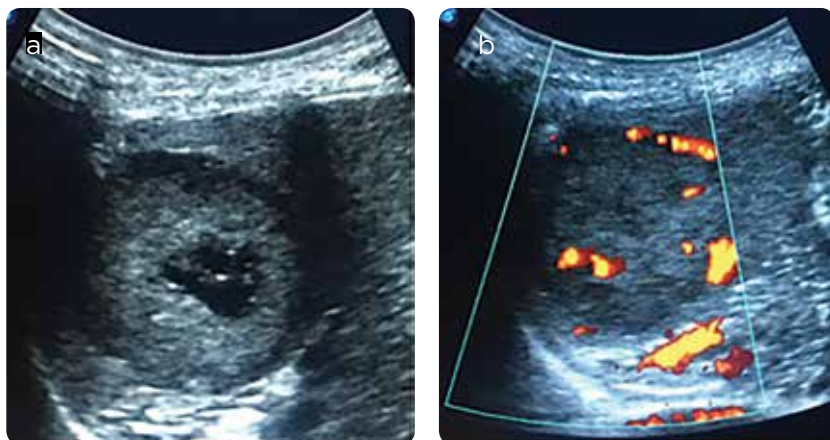


Figure 1. a) Abdominal ultrasound: mass of regular contours, isoechogenic to the parenchyma of the spleen, measures $4 \times 4,3 \times 4,2$ cm. Well-defined lesion of the surrounding splenic parenchyma, with an irregular peripheral hypo-echoic rim; internal anechoic region, representing necrosis. b) The color Doppler ultrasound shows a rich sign of blood flow in the portion of the tumor that appears solid. No blood flow was found in the anechoic region.

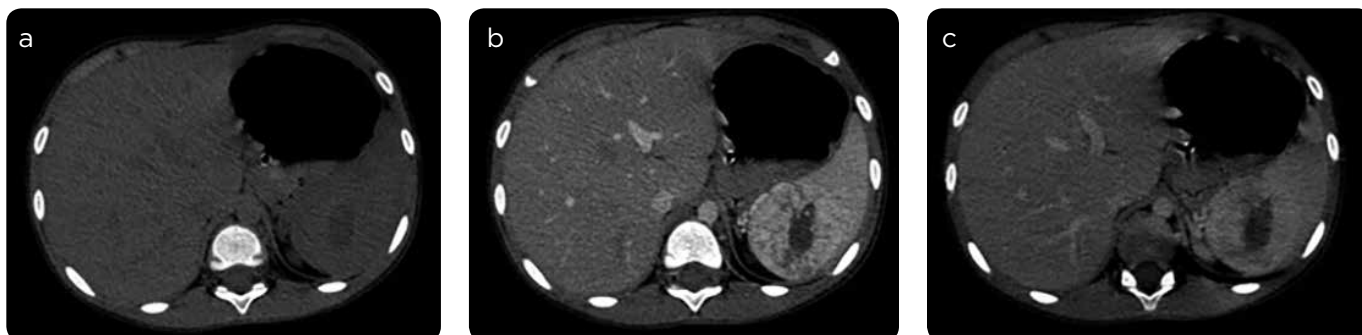


Figure 2. a) Single-phase CT scan: low signal zone in the upper pole of the spleen. b) Arterial phase: hypervascularized solid mass with strong heterogeneous enhancement, located in the upper pole and middle region of the spleen, with irregular edges, with necrotic center, measures $4,7 \times 4,3 \times 4,6$ cm. c) Venous phase: moderate lavage.

Laparoscopic splenectomy was performed in which the spleen was observed with a weight of 90 gr, measuring $9,5 \times 7,3$ cm, smooth capsule, shiny, covered by grayish surface.

For the pathological study, serial cuts were made that showed a solid nodule of $3,5 \times 3$ cm, greyish yellow with slightly lobed edges; the rest of the parenchyma was greyish brown, with poorly fixed areas. These findings are related to vascular neoplasia compatible with hemangioendothelioma.

Discussion

The spleen is a solid, highly vascularized organ located in the left upper quadrant in the most posterior region of the abdomen (6-8).

Histologically, the spleen is covered by a fibrous capsule containing afferent and efferent blood vessels, lymphatics and nerves. The red and white pulp can be distinguished in its parenchyma (7,8).

The red pulp is a network of vascular sinuses and terminal arterial branches that filters the blood. The white pulp has immune function. Macroscopically, it is an organized lymphoid tissue (Malpighi corpuscles) with T lymphocytes, some macrophages, and plasma cells (8).

In both ultrasound and CT, the spleen is usually isoechoic and isodense with the liver (6). In CT, the spleen is homogeneous, with an average attenuation between 40-60 HU, which is similar or less than in the normal liver (8).

After injection of contrast medium it may show heterogeneous enhancement in the arterial phase, of arciform and "brindle" presentation, due to differences in the flow of the red and white pulp, but it becomes

homogeneous approximately 60-90 seconds after administration of the contrast medium (6,7,9).

Splenic focal lesions may be tumoral (benign and malignant) or non-tumoral. The main benign tumour lesions are haemangiomas, hamartomas and lymphangiomas; malignant tumour lesions may be haematolymphoid or non-haematolymphoid.

The most important haematolymphoid lesions are non-Hodgkin's lymphomas and Hodgkin's disease. The main non-haematolymphoid lesions are angiosarcoma as primary tumor and metastases (6,7).

Splenic HE is a rare primary tumor of the spleen, which manifests as a vascular lesion with intermediate aggressiveness between benign hemangioma and highly malignant angiosarcoma (3,9-12).

Although its etiology is unknown, the average age is around 25 years, ranging from 3 to 54 years, but some pediatric cases have been reported; it has no gender predilection and the symptoms are abdominal pain, a palpable mass or hypersplenism (1,3,9).

The macroscopic pathological appearance of HE has been described as that of a solid, large, well circumscribed, non-encapsulated splenic mass. Its histological appearance can vary from well-differentiated patterns to highly undifferentiated forms, and the tumor is composed of vascular elements (1).

It is characterized by well-circumscribed lesions, properly separated from the healthy splenic parenchyma, moderate cellular atypia, little mitotic activity and an intermediate histological appearance between the cavernous hemangioma and conventional angiosarcoma (3). It typically shows a range of microscopic features with poorly defined vascular spaces, epithelioid morphology with mild cellular atypia, a low degree

of mitosis and no necrosis. Its malignancy potential is described as a border or intermediate line (9,10).

The absence of rapid growth and cellular atypia are the findings that distinguish HE from angiosarcoma (1).

Diagnosis remains a challenge, as the clinical features of HE are often nonspecific (1,10). With tumor growth, the patient may experience left upper abdominal pain, splenomegaly, and a palpable mass. In severe cases, symptoms include gastric distension, nausea, vomiting, dyspnea, male pain, and constipation (1). It has been described that in some cases chronic anaemia appears and there is a described case of splenic rupture (10).

On ultrasound, HE is typically seen as a hypoechogenic mass, distinct from the surrounding splenic parenchyma (4).

It may present anechoic areas that reflect intratumoral necrosis. In color Doppler images, HE appears with disordered vascularization, high velocity arterial flow, and a low resistive index in solid areas of the tumor. These findings are believed to correspond to tumor neoangiogenesis (1).

CT shows a low attenuation mass with increased solid portions of the tumor, which may appear hypovascular in relation to the normal splenic parenchyma.

Findings suggestive of malignancy, such as areas of necrosis and hemorrhage will not show enhancement. Signs of infiltration of the surrounding splenic parenchyma and evidence of metastatic disease can also be identified on CT. Calcification has not been described as a specific feature of this tumor.

On magnetic resonance imaging (MR), splenic HE appears as a solid, heterogeneous lesion with low signal in T1 and T2 weighted pulse sequences, an appearance suggesting hemosiderin (1).

CT is an important technique that helps to identify splenic HE; however, it is not possible to determine HE by images alone, since the findings are nonspecific (1,4), so pathological examination is required to arrive at the final diagnosis (1).

According to the histopathological type, HE is classified as epithelioid, retiform and kaposiform (3,10).

Hemangioendotelioma epitelioid: Occurs around medium and large caliber veins. Tumor cells are enlarged and often cuboid (similar to epithelial cells); in certain areas well-defined vascular ducts are barely visible. There is a proliferation of epithelial cells bulging to fusiform endothelial in nests or cords, in a fibromixoid stroma. The cytoplasm is vacuolated. The presence of pleomorphic nuclei and low mitotic activity is common. Cytokeratin is usually negative.

Retiform hemangioendotelioma: Occurs in young adults, without sex predilection. It shows a particular arboriform angiogenic growth similar to ridge nets, covered by embedded endothelial cells and monomorphs, with scarce cytoplasm and with minimal or no atypia. No atypical mitosis is observed. There is lymphocyte infiltrate that can occlude blood vessels, as well as being very close to endothelial cells, also small retiformed lesions, difficult to differentiate.

Kaposiform hemangioendotelioma: There are interconnected layers or nodules of fusiform endothelial cells lining the vessels in a cut or half-moon shape. Rounded vessels are also seen. Unlike Kaposi's sarcoma, nests of epithelioid-type endothelial cells can be found, with eosinophilic cytoplasm containing hemosiderin and vacuolated cytoplasm. Cell atypia is minimal and mitosis is infrequent (3,10).

Therapeutic strategies for splenic HE have been limited, as cases are rare. Complete splenectomy is considered the most effective therapy. However, this leads to increased postoperative risks, including the possibility of infection (1).

The prognosis is favorable even though it has a representative level of local infiltration. The risk of metastasis is low compared to that of recurrences (3).

Conclusion

Splenic HE is an infrequent pathology, but it has a good prognosis and no recurrences have been documented after fifteen years of its surgical removal. Diagnosis based on clinical and image alone is not possible, since the findings are non-specific, so the final diagnosis is histopathological.

Conflicts of interest: The authors declare that they have no conflict of interest.

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