

# Incidentally Discovered Asymptomatic Splenic Hamartoma with Rapidly Expansive Growth: A Case Report

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**Abstract.** *Splenic hamartoma (SH) is a very rare benign vascular lesion, usually asymptomatic. Although infrequent, it must be differentiated from malignant lesions, such as lymphoma or metastases, or other vascular neoplasms of the spleen. We present the case of a solid lesion of the spleen discovered incidentally in a 42-year-old woman, characterized by an unusual rapid expansive growth during four-month follow-up. The lesion, isoechoic and close to the hilum, was initially suspected to be an accessory spleen, measuring 3.5 cm in size. Four months later, magnetic resonance imaging revealed a 9-cm expansive nonhomogeneous mass in the antero-superior margin of the spleen, dislocating the stomach. The lesion exhibited central necrosis with hyper- and hypointense signal both in T1 and T2-weighted images. Due to the risk of spontaneous rupture and because malignancy could not be ruled out, the patient underwent hand-assisted laparoscopic splenectomy. Macroscopically, the spleen measured 15×12×4 cm and weighed 890 g. Cut sections revealed a single nodule of dark-red tissue sized 8.5 cm. The lesion exhibited a network of irregularly arranged and tortuous vascular channels lined by endothelium similar to splenic sinus lining cells, surrounded by aggregates of lymphocytes and macrophages resembling the pulp cords. On immunohistochemical staining the tissue was CD34 – and CD8 + and the final diagnosis was SH. In conclusion, when SH is suspected and malignancy cannot be ruled out, hand-assisted splenectomy should be considered the procedure of choice. Partial splenectomy should be preferred in children, to avoid potential risks of total splenectomy.*

Splenic hamartoma (SH) is a very rare benign vascular lesion, with an autopsy incidence of 0.02% and has only been documented in fewer than 200 cases (1-3). SH is usually asymptomatic, particularly in adults, but its association with myeloproliferative diseases and thrombocytopenia has been reported (4, 5). Histologically, SH comprises a disorganized overgrowth of mature cells and tissues that are normally present in the spleen, resulting in single or multiple nodules (6). Although infrequent, it must be differentiated from malignant lesions, such as lymphoma or metastases, or other vascular neoplasms of the spleen.

We present the case of an asymptomatic patient with a solid lesion of the spleen discovered incidentally, characterized by an unusual rapid expansive growth during four-month follow-up.

## Case Report

A splenic mass was incidentally found in a 42-year-old woman with a history of multiple sclerosis, who underwent abdominal ultrasound (US) due to right renal colic. The lesion, isoechoic and close to the hilum, was suspected to be an accessory spleen, measuring 3.5 cm in size. Physical examination and routine blood test were normal and no related symptoms were referred by the patient. Computed tomographic (CT) scan of the abdomen confirmed an isodense lesion of the spleen, characterized by low hyperdensity in the portal venous phase, which strengthened the suspicion of accessory spleen.

The patient was followed-up by US, showing a progressive and rapid enlargement of the lesion, without clinical or hematological alterations. Four months after the first US, magnetic resonance imaging (MRI) revealed a 9-cm expansive nonhomogeneous mass in the antero-superior margin of the spleen, dislocating the stomach. The lesion exhibited central necrosis with hyper- and hypointense signal both in T1- and T2-weighted images (Figure 1). The differential diagnosis was hemangioma, hamartoma or

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**Key Words:** Splenic hamartoma, splenic mass, splenic neoplasm, splenectomy.

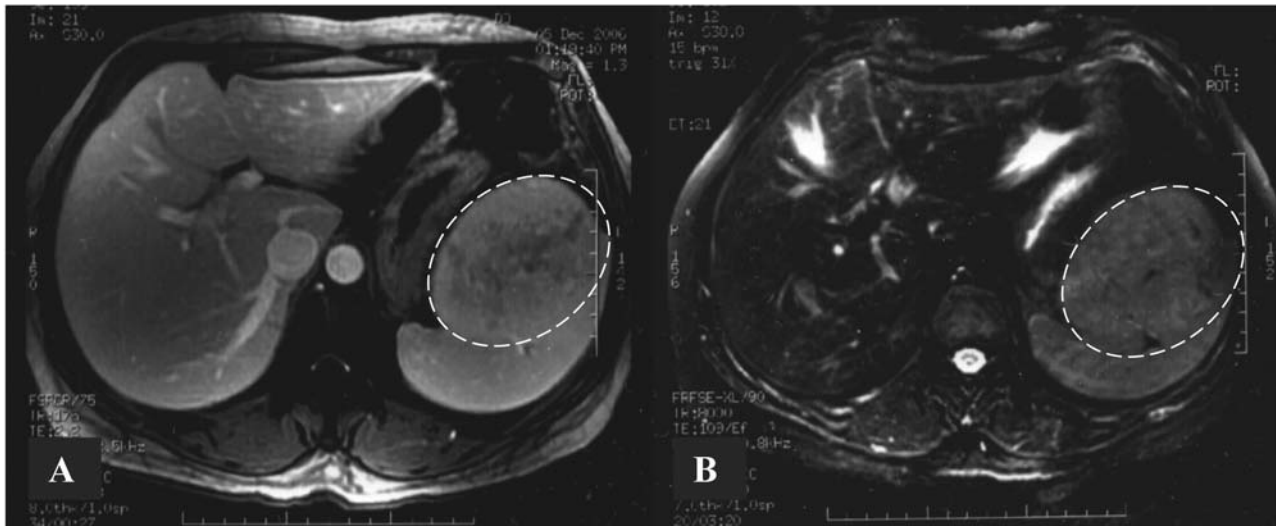


Figure 1. Magnetic resonance images showing a mass in the antero-superior margin of the spleen (dotted line), dislocating the stomach. The lesion exhibits central necrosis with hyper- and hypointense signal both in T1- (A) and T2-weighted (B) images.

metastasis. No sign of infiltration of other organs was found, but no clear signs of benignity were demonstrable.

Due to the risk of spontaneous rupture and because malignancy could not be ruled out, the patient underwent hand-assisted laparoscopic splenectomy. This approach preserves the advantages of laparoscopy (*i.e.* less postoperative pain and short hospital stay) but makes it possible to remove an intact organ, allowing a complete histological examination, avoiding both peritoneal dissemination in case of malignancy and fragmentation of the specimen (7).

Macroscopically, the removed spleen measured 15×12×4 cm and weighed 890 g. Cut sections revealed a single nodule of dark-red tissue sized 8.5 cm, with a similar color and consistency to that of the surrounding organ. The lesion was well demarcated from the normal splenic parenchyma, but without a capsule. On histological examination, an expansive nodule compressing the surrounding red pulp was observed. The lesion exhibited a network of irregularly arranged and tortuous vascular channels lined by endothelium similar to splenic sinus lining cells, surrounded by aggregates of lymphocytes and macrophages resembling the pulp cords. There were no organized lymphoid follicles, while foci of extramedullary hemopoiesis were observed. On immunohistochemical staining it was CD34 – and CD8 + and the final diagnosis was SH (Figure 2).

No complications developed during postoperative follow-up and the patient is asymptomatic 12 months after surgery.

## Discussion

Although vascular tumors are the most frequent neoplasm of the spleen, SH is an uncommon vascular lesion, firstly

described by Rokitansky in 1861 (8-11). SH, also called splenoma, is usually single and small, ranging between 2 and 7 cm in size and may occur at any age, with a mean age of 27 years for men and 37 years for women (9, 12). Reinterpretation and reclassification of these lesions revealed a novel, histologically unique entity, defined as sclerosing angiomatoid nodular transformation (SANT). This lesion usually appears as a multinodular, well-circumscribed mass containing a hypervascular core, frequently related to concurrent malignancies at other sites, such as lung or colonic cancer and leukemia (13).

SH is often discovered incidentally, during imaging studies performed for other reasons, or at autopsy (3, 14). Signs and symptoms can be associated with larger lesions, which typically occur in women, but symptomatic patients account for only 15% of cases (15, 16).

Unfortunately, the results of US and CT-scan are controversial and non-specific because SH does not have a characteristic radiologic appearance and thus, in several cases, malignancy cannot be excluded by imaging studies alone (6, 12). Moreover, the MRI pattern is heterogeneous and not definitive (6, 17). The atypical MRI appearance reported in our case, was previously described only in another report and referred to the presence of fibrous tissue in the lesion (18). Recently, a multi-modality imaging approach has been suggested, contributing to improving the diagnostic accuracy (3). Fine-needle aspiration cytology is rarely performed and cases of challenging cytological diagnosis have been reported (6, 19).

Histologically, it may be difficult to differentiate SH from capillary hemangioma or other vascular lesions of the spleen and immunohistochemical staining is required to

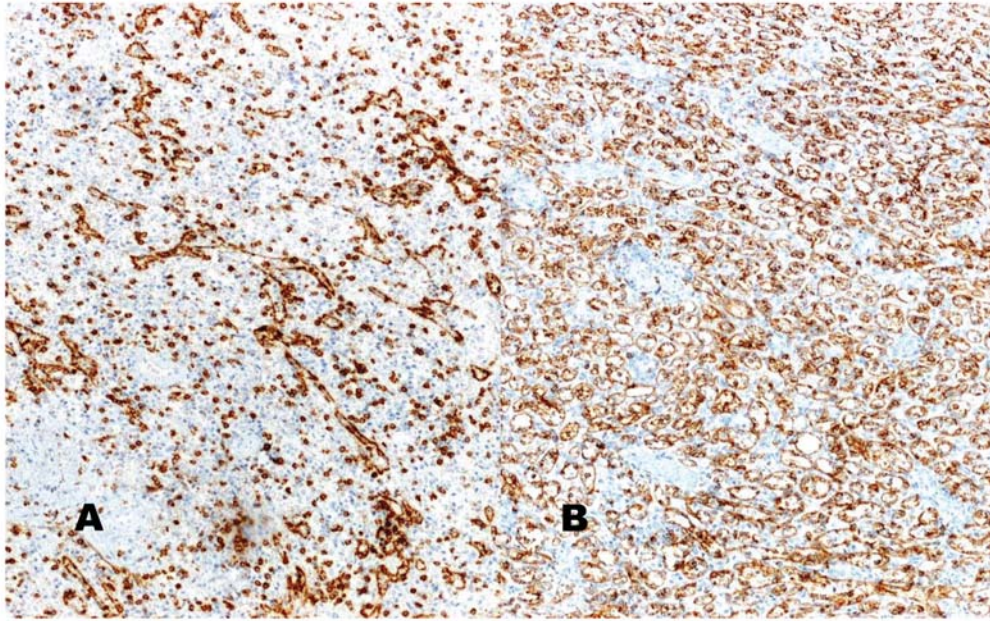


Figure 2. A: Normal spleen (hematoxylin-eosin stain). B: Splenic hamartoma, with positive immunohistochemical stain for CD8. Original magnification,  $\times 40$ .

confirm the diagnosis (20). SH contains sinus and pulp cordlike elements, whereas cord capillary hemangiomas encompass well-organized lymphoid tissue (21, 22). Because of its origin from splenic sinusoids, SH exhibits reactivity for CD8 and factor VIII-related antigen and negativity for CD34 (23, 24). The endothelial cells of SH are CD8 +, whereas those of hemangioma are not and thus SHs have a characteristic CD34 –/CD21 –/CD8 + phenotype (24).

When a SH is suspected and malignancy cannot be ruled out, elective splenectomy should be considered. Furthermore, hematological disorders associated with SH are related to hypersplenism and can be cured by splenectomy. Only a few cases of spontaneous rupture have been reported, mainly related to coexisting diseases such as liver cirrhosis and portal hypertension, representing a life-threatening complication (25, 26). However, in our case, the rapid growth of the lesion suggested the need for splenectomy. Alternatively, the age of the patient, size of the mass and results of imaging studies might indicate to splenic preservation or partial resection of the spleen.

In conclusion, in patients with SH, hand-assisted laparoscopic splenectomy is the procedure of choice in the presence of a suspicious mass. Partial splenectomy should be preferred in children, to avoid potential risks of total splenectomy. In any case, immunohistochemical studies are needed to obtain the final diagnosis of any enlarging mass of the spleen.

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