

## Symptomatic splenoma (hamartoma) of the spleen. A case report

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### Abstract

Hamartomas of the spleen (splenomas) are very rare benign tumors composed of an aberrant mixture of normal splenic elements. Herein we present a unique case of a symptomatic non-palpable splenoma in a 64-year-old female patient presented with anemia and thrombocytopenia and we describe imaging findings in ultrasound, computed tomography and magnetic resonance imaging. To our knowledge, this is the first case of a relatively small splenic hamartoma (35 mm at histopathology) associated with thrombocytopenia and anemia that resolved completely several months after splenectomy. Hippokratia 2010; 14 (1): 54-56

**Key words:** spleen, splenoma, hamartoma

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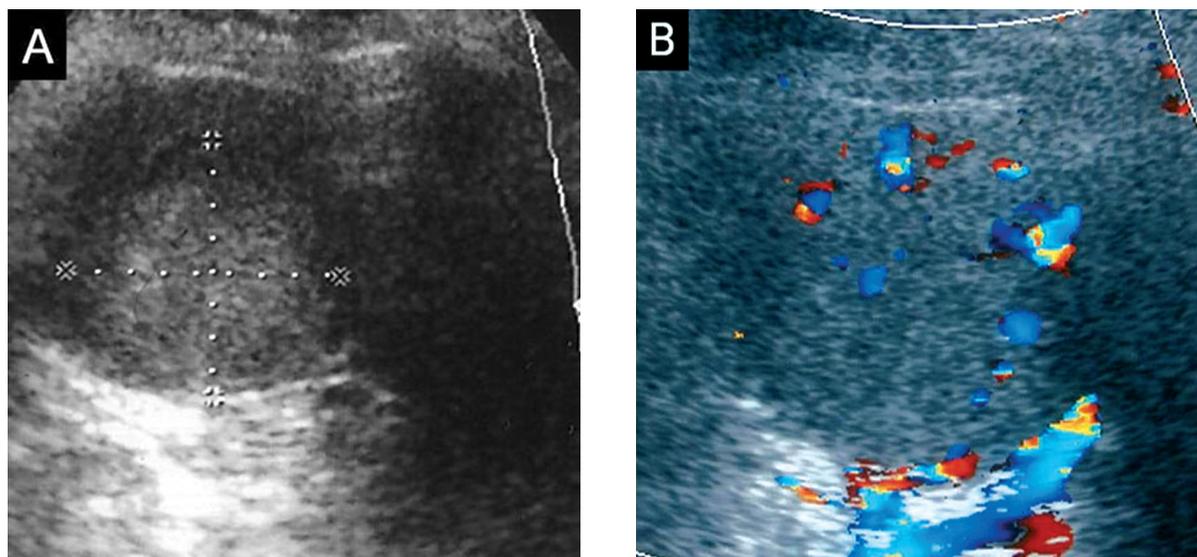
Hamartoma of the spleen (splenoma) is a rare benign tumor composed of an aberrant mixture of normal spleen tissue<sup>1-3</sup>. Herein, we present a case of a relatively small (35 mm at histopathology), non-palpable symptomatic splenoma in a 64-year-old female patient presented with anemia and thrombocytopenia that resolved completely after splenectomy.

### Case report

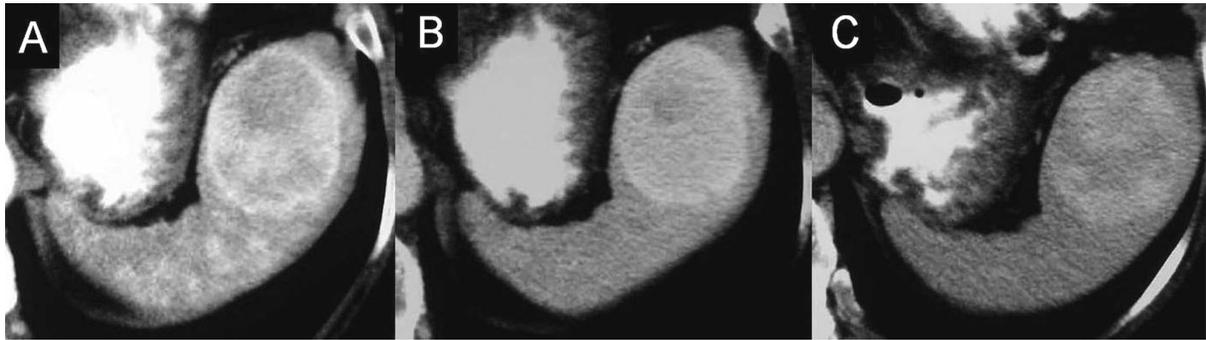
A 64-year-old female patient with free past medical history was referred to our department for abdominal ultrasound (US) during investigation of anemia and throm-

bocytopenia. US revealed a solid, slightly hyperechoic mass in the upper pole of the spleen with a maximal diameter of 51 mm, with sharp borders and with internal vascularization on Power Doppler (Figure 1).

Further investigation with triphasic helical computed tomography (CT) was performed the same day. During arterial phase the lesion demonstrated intense inhomogeneous enhancement, similar to splenic parenchyma, with a peripheral enhancing rim. In portal phase the lesion was more hyperdense than splenic parenchyma, while in the delayed phase, the lesion demonstrated delayed 'wash out' (Figure 2).



**Figure 1:** US of the splenic lesion, demonstrating a well defined hyperechoic lesion in the upper pole of the spleen (A) with increased internal vascularization in Power Doppler (B).



**Figure 2:** Triphasic spiral CT demonstrating the lesion with intense, inhomogeneous enhancement during arterial phase (A). In portal phase the lesion is hyperdense compared to normal splenic parenchyma (B). Delayed “wash out” is demonstrated in the delayed phase (C).

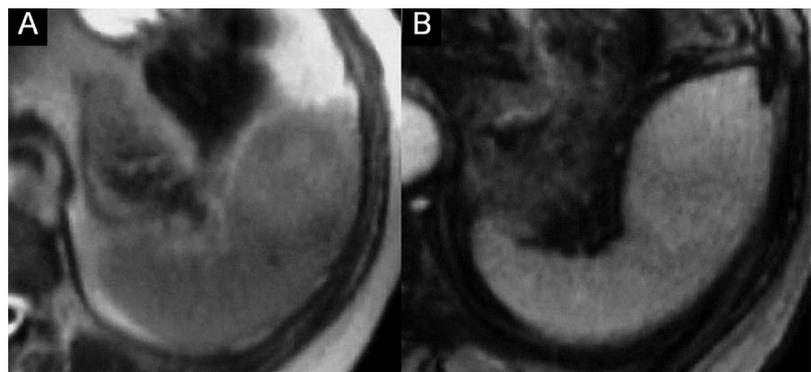
Magnetic resonance imaging (MRI) of the abdomen was performed the next day. The lesion demonstrated intermediate to high signal intensity in T2-Haste images comparative to normal splenic parenchyma, with homogenous intense enhancement after administration of gadolinium (Figure 3).

Splenectomy was finally performed a week later because of persistent patient's symptoms. Histopathological evaluation confirmed the diagnosis of a splenoma with greatest diameter of 35 mm, with the tumor consisting exclusively from red pulp that was positive in CD-31, CD-34 and CD-45 RO immunohistochemical strains (Figure 4). The rest of the splenic parenchyma was normal.

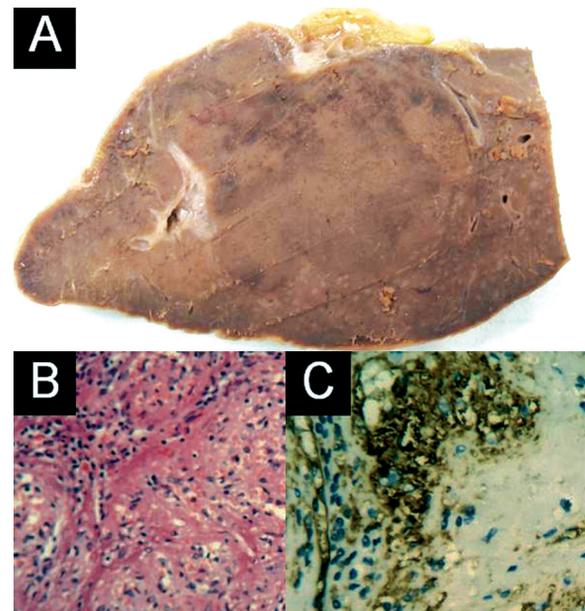
Finally, the patient was discharged from the hospital on the 12th postoperative day. Blood counts returned to normal values 4 months later, and 24 months after, the patient is still asymptomatic, in general good condition and with normal blood counts.

### Discussion

Splenoma is a rare benign tumor which consists of aberrant splenic tissue<sup>1</sup>. It was first described in 1861 by Rokitansky and since then about 140 cases have been reported. Its frequency in autopsy series is reported to be 0,024-0,13%<sup>2</sup>. Splenomas are usually discovered incidentally in all ages with supremacy in elderly women. Although splenomas are usually asymptomatic, they may rarely cause symptoms due to splenomegaly (a feeling of weight in the left upper quadrant, splenic rupture) or due to hypersplenism (anemia, thrombocytopenia)<sup>3</sup>. They may coexist with other hamartomas in other organs, with tuberous sclerosis and with Wiskott - Aldrich - like syndrome. A relationship with other neoplastic diseases has been also reported<sup>4,5</sup>. To our knowledge, this is the first described case of a relatively small splenoma (35 mm at



**Figure 3:** MRI of the lesion, with increased signal intensity relatively to the normal splenic parenchyma on axial T2-Haste images (A) and heterogeneous enhancement on axial T1-Flash images after intravenous administration of gadolinium (B).



**Figure 4:** A. Surgical specimen showing a well circumscribed lesion with a maximum diameter of 35mm in the spleen. B. Histological section showing the cells of red pulp of the lesion (HE X 200). C. Immunohistochemical staining showing the cells to react with factor CD-31.

histopathology) in an adult patient causing signs of hypersplenism (anemia and thrombocytopenia). In adults, reported symptomatic splenomas that manifested with signs of hypersplenism (anemia, thrombocytopenia) were larger than 9 cm<sup>6,7</sup>. In children, solitary splenomas causing signs of hypersplenism were larger than 4 cm (4-18cm). There are also reported cases of children presented with signs of hypersplenism with multiple splenomas, ranging from 2-5cm<sup>3,8-11</sup>.

Splenomas are well circumscribed, solid nodular lesions that compress the splenic parenchyma without infiltrating it. They are usually solitary tumors that rarely contain calcifications or grow to huge dimensions. A splenoma with a diameter of 19 cm has been reported<sup>12</sup>. Histologically 3 types of splenoma have been described. Type I develops from the white pulp and consists of abnormal lymphoid tissue. Type II develops from red pulp and consists of abnormal sinuses complex. Type III which is the most common type, is a combination of types I and II and contains elements of both types<sup>6,13</sup>.

Imaging findings of splenomas are not specific and depend on their type. On US, splenomas are usually solid, homogenous lesions with sharp borders. They can rarely demonstrate cystic degeneration and calcifications, which are due to ischaemia or hemorrhage. They are usually hyperechoic related to the normal splenic parenchyma with intense vascularization on Color Doppler<sup>14</sup>. Only one case of avascular splenoma has been reported, that showed large amorphous calcifications<sup>15</sup>. On CT, splenomas are usually isodense to normal splenic parenchyma, before and after intravenous administration of contrast media. Usually, the only finding is an abnormality of the splenic contour, with no signs of invasion. An intense heterogeneous enhancement in the arterial phase and on delayed images like in our case has also been reported<sup>7</sup>. On MRI splenomas are isointense to splenic parenchyma on T1 weighted images and hyperintense on T2 weighted images. After gadolinium administration, they usually demonstrate heterogeneous enhancement<sup>16</sup>.

Radiological differential diagnosis of splenoma should include solid mass-forming lesions of the spleen such as lymphoma, metastatic lesions, inflammatory myofibroblastic tumor, disseminated fungal or mycobacterial infections, sarcoidosis and vascular tumors of the spleen, including hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, sclerosing angiomatoid nodular transformation of the spleen and angiosarcoma. Immunohistochemically the lining cells of the vascular channels of the splenoma are positive for endothelial markers CD-8, CD-31, factor VIII-related antigen and vimentin and negative for endothelial markers CD-21 and

CD-68, although endothelial marker CD-68 is positive in scattered stromal macrophages<sup>13</sup>.

In conclusion, although splenoma is a very rare tumor, it must be included in the differential diagnosis of splenic lesions. To our knowledge, this is the first case of a relatively small splenoma associated with thrombocytopenia and anemia that resolved completely a few months after splenectomy. Based on patient's history and follow up, we assume that splenoma, although small, demonstrated abnormal splenic over-functionality, mimicking hypersplenism, since spleen size and histology were normal and blood counts returned to normal after splenectomy.

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