Splenic hamartoma in a patient with peripancreatic arterial arcades: A case report

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ABSTRACT
Splenic hamartoma is a rare benign vascular lesion of the spleen. A splenic mass was incidentally detected in an asymptomatic 65-year-old male during an abdominal ultrasound scan. The workup of the splenic lesion included an abdominal CT angiography (CTA), contrast-enhanced ultrasound (CEUS), and microvascular flow imaging Doppler sonography. The CT scan confirmed that the splenic mass had benign characteristics, and its contrast enhancement was similar to the spleen. The CEUS was able to rule out the possibility of hemangioma. Meanwhile, chronic compression of the celiac trunk and the filling of the splenic artery from collaterals were also detected on CTA. The imaging studies suggested a splenic hamartoma, and the diagnosis was confirmed by the result of a fine needle aspiration biopsy.

KEYWORDS
splenic hamartoma, spleen tumor, splenoma, FNAB

Introduction
Splenic hamartoma is a rare benign vascular proliferative neoplasm of the vascular endothelial lining cells [1]. They are often present as a solitary mass, but multiple lesions have been reported [2].

Most patients are asymptomatic and splenic hamartomas are usually identified incidentally on imaging [3]. The most frequent cause of chronic celiac trunk obstruction is compression by the median arcuate ligament (MAL), which is also called Dunbar syndrome [4]. MAL syndrome is a rare cause of splenic infarcts, but it has not been associated with splenic hamartoma previously.

Contrast-enhanced ultrasound (CEUS) is routinely used for the detection and characterization of splenic lesions. The spleen shares the property of sequestering and retaining ultrasound contrast microbubbles with the liver and is also ideally suited for ultrasound contrast imaging [5]. Microvascular flow imaging (MVFI) is an advanced Doppler ultrasound technique to visualize slow velocity flow in small-caliber vessels [6]. This is the first report on CEUS and MVFI aiding the diagnosis of a splenic hamartoma.

Case report
A splenic mass was incidentally found in a 65-year-old man with a history of prediabetes, parathyroid adenoma, hyperparathyroidism-jaw tumor syndrome, and gout. During an
abdominal ultrasound examination, an 80 × 90 mm mass was depicted between the diaphragm and the spleen, which could not be demarcated from the splenic surface.

A multiphase contrast-enhanced abdominal computed tomography (CT) (Fig. 1) revealed an 80 × 90 mm dumbbell shape lesion protruding from the subphrenic contour and the hilar surface of the spleen. The lesion was isodense to the spleen in the unenhanced scan, displayed a few prominent arteries around its surface, and enhanced identical to the splenic parenchyma in the portal venous and delayed phases except the hypovascular center. The CT morphology raised the possibility of a splenic hamartoma or hemangioma. The CT angiography (contrast agent: 60 ml Ultravist 370, Schering, Germany) scan also showed chronic compression of the celiac trunk at its origin by the thickened MAL, a peripancreatic network of collateral arteries, and retrograde filling of the splenic artery from the gastroduodenal and the dorsal pancreatic arteries.

The splenic lesion was further characterized by CEUS and MVFI. The mass was nearly isoechoic with the exception of a slightly hyperechoic center on B-mode ultrasound (Fig. 2/A) scanned with a Samsung RS85 Prestige ultrasound system (Samsung Medison Co. Ltd., Hongcheon, Korea) using the CA 1-7S convex probe with general preset by an expert radiologist with more than 10 years of experience in abdominal ultrasonography. The lesion showed irregular-shaped vascular trunks at its periphery, otherwise, it was indistinguishable from the rest of the spleen on MVFI (Fig. 2/B). After the injection of the microbubble contrast agent (1.5 ml in bolus SonoVue, Bracco, Milan, Italy), arterial then venous phase data collection was performed for 4 min during dynamic testing. The lesion showed peripheral enhancement similar to the normal spleen with no contrast accumulation in the central part (Fig. 2/C and D). The enhancing area did not show contrast washout. The CEUS ruled out the possibility of a hemangioma.

The final diagnosis was established with an ultrasound-guided fine-needle aspiration biopsy (FNAB) (Fig. 2/E). The cytology examination showed leached red blood cells in a thin eosinophilic background, on cell-rich smears, fragments composed of mixed lymphoid cells arranged in cell groups around blood vessels could be seen (Fig. 2/E). In addition, dissociated cellular elements showing lymphocytes, plasma cells, and neutrophil granulocytes appeared. The cytopathology findings were consistent with the splenic hamartoma or splenoma diagnosis and ruled out malignancy.

**Discussion**

Splenic hamartomas were first reported by Rokitansky in 1861. They are non-capsetulated, single or multiple nodules in the spleen and consist of grossly disproportionate native splenic elements [3]. However vascular tumors are frequent lesions of the spleen, splenic hamartoma is rare. Only around 200 cases have been reported [2].

Splenomas are usually incidental findings with an incidence of 0.024%–0.13% [7]. More than 80% of the cases are asymptomatic, and splenic hamartomas are found normally during imaging, surgery, or postmortem [1]. Splenomas may also cause anemia, thrombocytopenia, or pancytopenia. Other less common symptoms include fever, night sweats, malaise, and spontaneous rupture [7]. This clinical symptomatology seems to occur more often in adults, especially in women [3].

Although splenomas are rare, they must be differentiated from other vascular lesions like spleen tumors. Solid mass-forming lesions, such as metastases and lymphoma, mycobacterial infections, and sarcoidosis must be included in the radiological differential diagnosis [8].

The pathogenesis of splenic hamartoma is debated; a congenital malformation, a disorganized growth of the red pulp, a neoplastic proliferation, and a reactive lesion to prior trauma have been considered as potential explanations of its origin [9]. In our case, there is a possibility that the splenic hamartoma has been a reactive lesion caused by the altered blood flow in the splenic artery and perisplenic collateral arteries because of the chronic celiac trunk compression.
Fig. 2. Ultrasonography (B-mode, MVFI, CEUS) and Cytopathology (FNAB). A: An 80 × 90 mm isoechoic mass with a slightly hyperechoic center was detected on B-mode ultrasound. B: The splenic mass displayed a microvascular flow pattern indistinguishable from the rest of the spleen with MVFI. C: The lesion showed enhancement dynamics similar to the normal spleen with CEUS. D: B-mode reference image. E: Fine-needle aspiration biopsy of the splenoma. F: Thick tissue fragments composed of mixed lymphoid cells arranged in cell groups around blood vessels could be seen.
Due to his hyperparathyroidism-jaw tumor syndrome, the patient also has a higher risk of tumors.

With more frequent use of modern imaging techniques, the incidence of accidentally detected splenic lesions is also increasing. Recently, Wang et al. [10] described the radiological features of splenic hamartomas. Hamartomas typically appear as a single well-defined homogeneous isoechoic to hyperechoic nodule or mass on ultrasound.

On CT, hamartomas appear as solid single or multiple masses that are well-circumscribed and encapsulated. They are slightly hypodense before contrast administration. After contrast injection, prolonged enhancement is achieved in a single mass, although low-density masses are observed in multiple splenic hamartomas [8, 10].

The use of CEUS frequently provides valuable additional information to narrow the differential diagnosis and particularly to triage lesions that are likely to be benign from those that may be malignant [5]. The MVFI is capable of real-time, highly detailed visualization of small velocity vessels without a contrast agent. MVFI uses advanced multidirectional filters, which can separate microvascular flow from tissue clutter by analyzing the signal’s spatiotemporal coherence [6].

Both CEUS and MVFI were able to exclude other common vascular proliferative lesions such as hemangioma, sclerosing angiomatous malformation, and angiosarcoma from differential diagnosis by demonstrating a contrast enhancement dynamics and a microvessel distribution pattern similar to the normal spleen.

Conclusion

Although splenic hamartomas are very rare, they must be included in the differential diagnosis of splenic lesions. Using multimodal radiologic imaging can be useful to identify the diagnosis. The definitive diagnosis depends on pathologic examination, with the main purpose of differentiating this benign lesion from a malignant tumor.

Conflicts of Interest: One of the co-authors, Dr. Viktor Bérczi, is the Deputy Editor-in-Chief, the corresponding author, Dr. Pál Kaposi is an associate editor of IMAGING, therefore the submission was handled by a different member of the editorial team.

REFERENCES


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