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Case Report

Primary splenic lymphoma on top of intrahepatic splenosis: A unique case report☆,☆☆,★

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ABSTRACT

Solid organ splenosis is a challenging diagnosis with many atypical imaging features that can overlap with neoplastic masses of the affected organ. We present a sporadic case of intrahepatic splenosis in a 68-year-old woman with transformation into a low-grade B cell lymphoma. Initial cross-sectional imaging suggested focal nodular hyperplasia (FNH) ruled out on contrast-enhanced Magnetic Resonance Imaging (MRI) using a hepatobiliary-specific contrast agent. A Tc-99m sulfur colloid scan was negative. The final diagnosis was confirmed by a needle-guided biopsy revealing intrahepatic splenosis with transformation into a low-grade B cell lymphoma.

Introduction

Splenosis is an acquired benign condition that may follow splenectomy or traumatic splenic injury [1]. Theoretically, splenosis occurs due to spillage of splenic sinusoidal cells that are directly implanted into different compartments of the peritoneal cavity or even into the thoracic cavity if the diaphragm was breached [1]. Distant implantation of heterotopic splenic tissue can occur by hematogenous spread to different organs such as the liver, skin, and brain [2].

Intrahepatic splenosis a condition in which splenic sinusoidal tissue embeds into the hepatic parenchyma. This rare entity was first reported in the literature in 1939 Since then, 59 cases have been reported [3,4]. A fibrous capsule usually surrounds the splenic tissue, thus supporting the hematogenous implantation theory. The imaging characteristics of these lesions are atypical and often misdiagnosed

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with subsequent unnecessary biopsies or surgical interventions [5]. To our knowledge, we are reporting the first case of intrahepatic splenosis transforming into primary splenic low-grade B-cell lymphoma. Additionally, we review the role of different imaging modalities in differentiating intrahepatic splenosis from other hepatic focal lesions.

Case report

A 64-year-old woman presented with persistent weakness, fatigue, anxiety, and occasional abdominal pain. She had a history of post-traumatic splenectomy 30 years ago. The patient was first encountered with diffuse non–radiating abdominal pain in the emergency department. Her family and social history were noncontributory. A review of other systems revealed no significant abnormality.

On physical examination, the patient was stable, and not in acute distress. Vital signs, including orthostatic, were normal. Abdominal examination revealed a surgical abdominal scar of previous laparotomy and mild tenderness on palpation. However, there was no guarding nor rigidity. Laboratory studies were unremarkable.

A computed tomography (CT) study of the abdomen and pelvis was requested to further assess the patient's complaint. On contrast-enhanced CT, a 2.1×1.1 cm soft tissue nodule in the left upper quadrant was favored to be a residual splenic tissue. Furthermore, there were 2 indeterminate hyperattenuating masses within the right lobe of the liver measuring up to 3.5 cm, and 1.6 cm (Fig. 1). The patient refused to receive MRI contrast material, so further evaluation with non–contrast MRI was performed. It was also inconclusive, and the diagnosis of focal nodular hyperplasia (FNH) was suggested based on the MRI and contrast-enhanced CT findings. After discussing the diagnostic possibilities with the patient, another follow-up MRI study using a hepatobiliary contrast agent (Gadoxetate-Disodium) was performed to characterize the liver lesions better. The liver lesions were hypointense on T1-WIs, hyperintense on T2-WIs, and showed heterogeneous arterial and portal phases enhancement. The lesions did not retain the contrast on the delayed 20 minutes post-contrast images, making the diagnosis of FNH improbable (Figs. 2 and 3). Given the patient's history of splenectomy, the rare diagnosis of intrahepatic splenosis was considered, and correlation with scintigraphic imaging was recommended.

Tc-99m Sulfur Colloid SPECT-CT scan showed increased radiotracer uptake within the left upper quadrant soft tissue consistent with residual or hypertrophied splenic tissue. There was no tracer uptake by the 2 focal hepatic lesions, making the diagnosis of splenosis less likely (Fig. 4). Given the negative sulfur colloid imaging, a tissue biopsy was performed. Histologic examination revealed scattered neoplastic B cells within the splenic sinusoids and perivascular areas—a characteristic picture of low-grade B cell neoplasm within intrahepatic splenosis (Fig. 5). There was no lymphadenopathy elsewhere on CT imaging of the chest, abdomen, and pelvis. Bone marrow biopsy showed no marrow involvement. Additionally, the CBC revealed no blast cells. Given the indolent nature of the disease, the patient received no treatment,

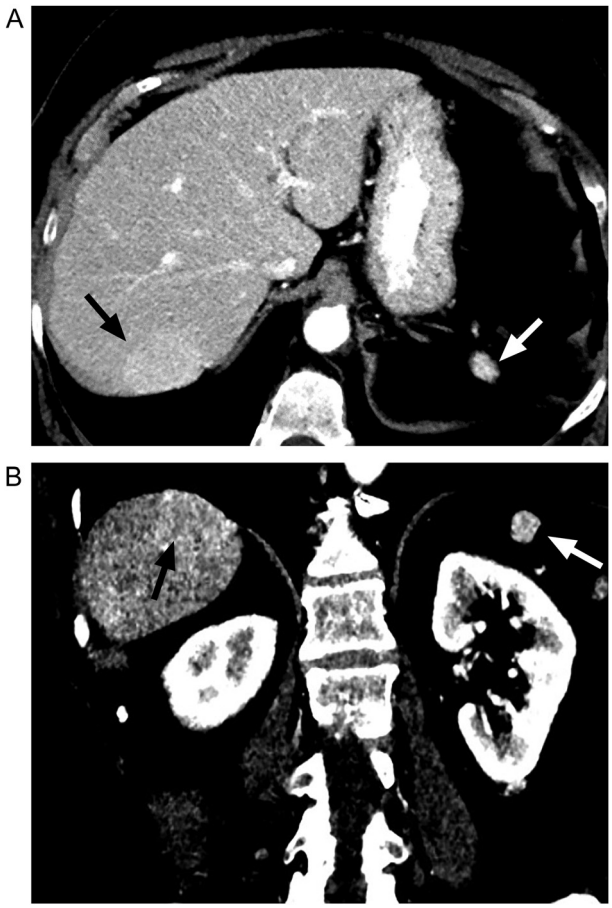

Fig. 1 – Contrast-enhanced axial (A) and coronal (B) CT images show sub-capsular enhancing lesion in the right hepatic lobe (black arrows). Additionally, a left upper quadrant small soft tissue density (white arrow) is present, likely residual splenic tissue.

and was required to get the continued follow-up. The hepatic lesions remained stable in size over 2 years of follow-up imaging.

Discussion

B-cell lymphoma represents the majority of non–Hodgkin lymphoma (NHL), which is related to a malignant proliferation of B cell progenitors. NHL is either high- or low-grade neoplasia according to histologic differentiation [6]. NHL can be classified according to their behavior into indolent, aggressive, and highly aggressive lymphomas. While low-grade NHL is more indolent, high-grade ones are more aggressive with a predominance of blast cells with a higher mitotic rate. Moreover, aggressive lymphomas frequently affect the hematopoietic precursors early in the course of the disease [7]. On the other hand, indolent lymphomas have a more benign course and better response to therapy, although their clinical course can be marked by multiple periods of remission, and relapse. Clinical manifestations of low-grade

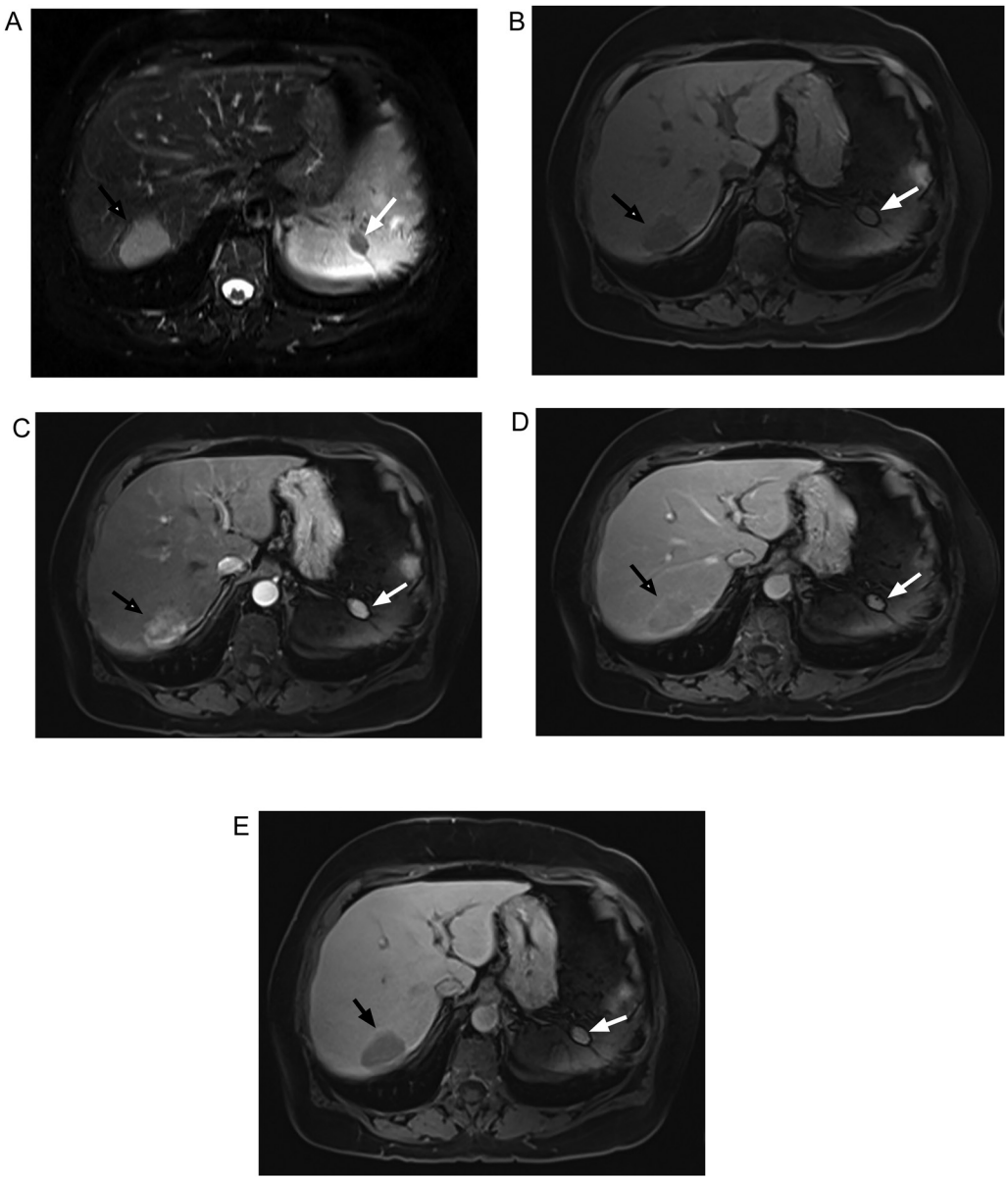

Fig. 2 – (A) Axial T2 TSE FS, (B) Pre-contrast T1 FS, and (C- E) dynamic post Gadoxetate disodium injection. The images demonstrate a T2 hyperintense and T1 hypointense hepatic focal lesion in segment VII (black arrows). The lesion elicits heterogeneous arterial enhancement, hypo-enhancement relative to liver parenchyma on portal-venous phase. No retention of contrast on delayed (20 minutes) imaging. The left upper quadrant small soft tissue nodule (remnant splenic tissue) has similar signal characteristics to the liver lesion (white arrows).

NHL are non–specific, such as weight loss, weakness, fatigue, and enlarged spleen [8]. As a lymphoid organ, the spleen can be secondarily involved in lymphomas. However, primary splenic lymphoma is a rare condition with an estimated incidence of 1%. The definitive diagnosis of primary splenic lymphoma is made when the disease is limited to the spleen without extra splenic involvement [9].

Splenosis is defined as heterotopic implantation of splenic tissue [10]. It is observed in more than 67% of patients after splenic rupture. Intrahepatic implantation of splenic tissue is rare and occurs when splenic sinusoidal cells are implanted into the liver after a splenic injury. The previously reported cases of intrahepatic splenosis were all diagnosed incidentally, as in our case. Although most patients are asymptomatic, few cases presented with diarrhea, pain, and bowel obstruction from adhesions, with an imaging study showing an indeterminant hepatic lesion. Depending on the age and underlying existing liver disease, those lesions were mistakenly diagnosed as hepatocellular carcinoma (HCC), [11] hepatic adenoma, [5] or FNH as in our case. Moreover, the final diagnosis was only possible at histologic examination [12]. A false diagnosis such as HCC and hepatic adenoma may lead to unnecessary treatment [10,11]. The CT findings of splenosis are non–specific. In our case, both lesions were described as

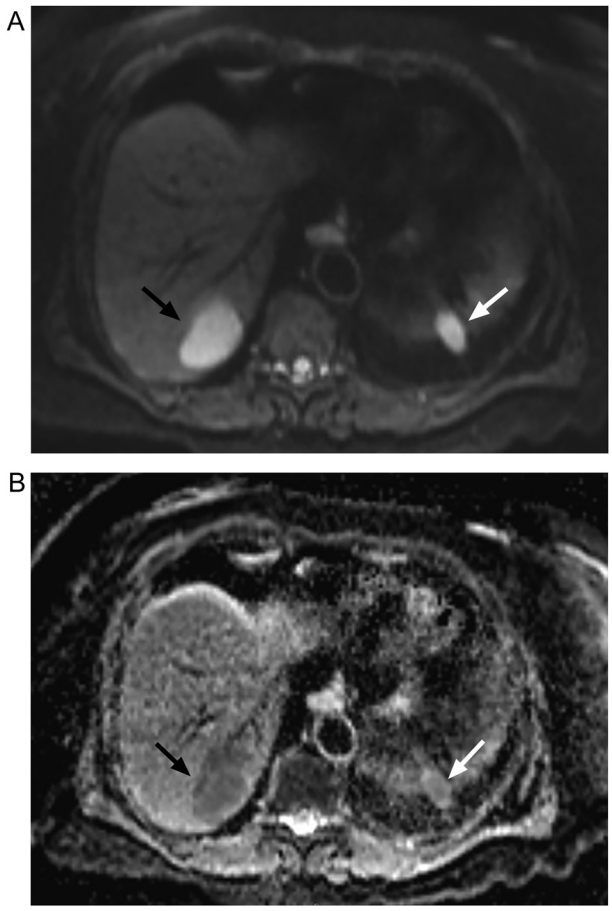

Fig. 3 – Axial DWI (Diffusion-Weighted Imaging) b = 500 (A) and corresponding ADC (Apparent Diffusion Coefficient) map (B) images show a subdiaphragmatic hepatic focal lesion with high signal intensity on DWI and restricted diffusion on ADC (black arrows). The left upper quadrant soft tissue nodule has similar diffusion pattern (white arrows).

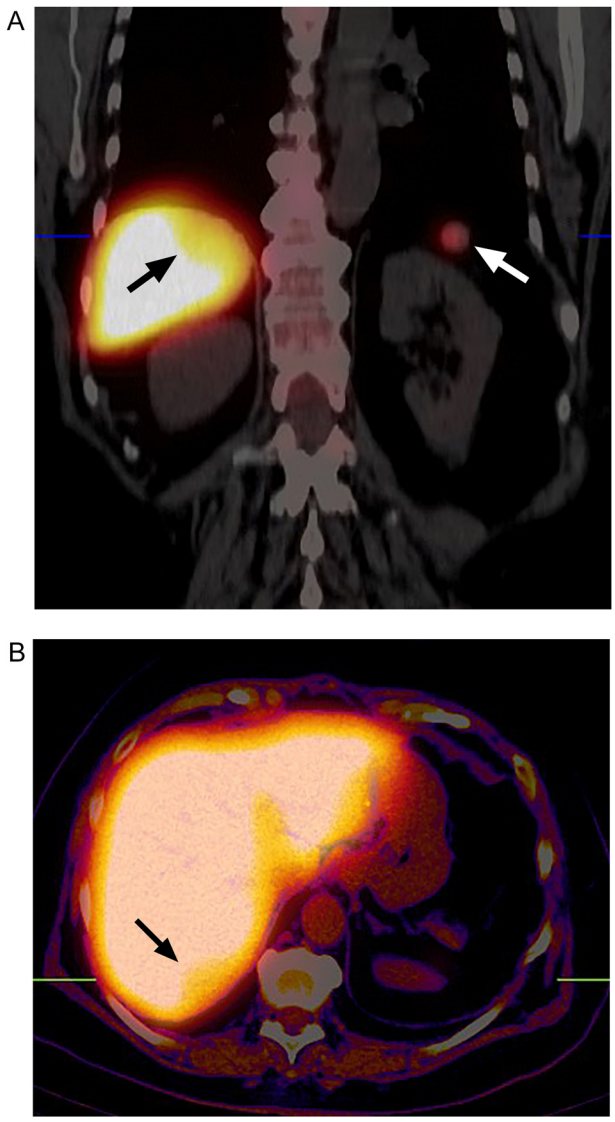

Fig. 4 – (A) Coronal and (B) axial Tc-99m sulfur colloid fused SPECT/CT images showing a small splenic tissue uptake in the left upper quadrant (white arrow). No radiotracer uptake was noted within the intrahepatic lesion (black arrows).

enhancing intrahepatic lesions that could not be further characterized (Fig. 1) [12]. Ultrasound features are also nonspecific. On MRI, splenosis is usually described as low T1 signal intensity, iso to high signal intensity on T2 weighted images, and shows heterogeneous enhancement on arterial phase similar to the signal and enhancement patterns of the spleen usually described as geographic or zebra pattern of enhancement [13]. The presence of a thin hypointense fibrous capsule can sometimes be noted [1]. Diffusion-Weighted Imaging (DWI) may help diagnose splenosis. Splenic tissue yields the most restriction in diffusion among other abdominal organs. However, this is also a non–specific finding that may overlap with other solid hepatic tumors. Special contrast agents could differentiate between hepatic splenosis and hepatic neoplasms. Superparamagnetic iron oxide (SPIO) contrast MRI can be used in such cases. This contrast agent is taken up by splenic tissue, causing a significant decrease in signal intensity of the splenic tissues on post-contrast images [1]. In light of the patient's history of the previous splenectomy, the diagnosis of splenosis was more favorable. A scintigraphic study was ordered to confirm the preliminary diagnosis. There are 2 dedicated scintigraphic methods for detecting heterotopic splenic tissue, including Tc-99m-labeled heat-denatured red blood cells (HRBC), and Tc-99m sulfur colloid scans. Although Tc-99m-labeled (HRBC) is more sensitive due to 90% sequestration into splenic tissue, it is more difficult to apply and not readily available in every institution. It is also very operator dependent. Tc-99m sulfur colloid is taken up by the reticuloendothelial system and shows more than 80% hepatic uptake and only 5%-10% splenic uptake [14]. Although small size limits the sensitivity, using SPECT/CT can make up for such drawbacks by increasing the overall sensitivity of splenic tissue detection [15].

Intrahepatic splenosis is a rare incidental condition, and diagnosis can be difficult when there are no typical imaging features. Although the MRI and dedicated nuclear medicine

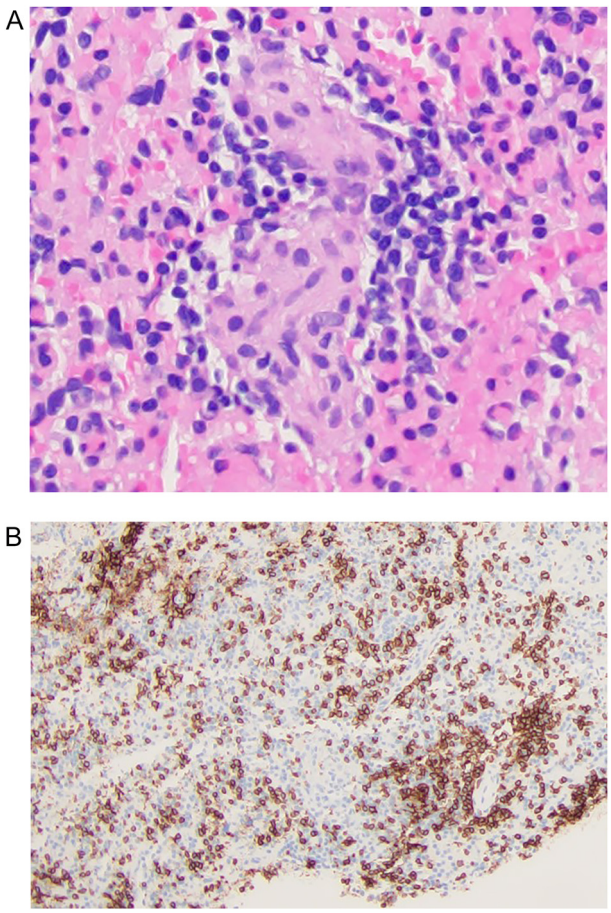

Fig. 5 – (A) H&E slide (original magnification x200) shows small lymphocytes within the sinusoids and perivascular areas representing lymphocytic infiltration of splenic tissue. (B) Immunohistochemical stain for CD20 (original magnification x 100) staining the small lymphocytes in brown color (Color version of the figure is available online.)

imaging are sensitive and specific for diagnosis, Tc-99m sulfur colloid imaging was false negative in our case, likely due to the lymphomatous transformation of the intrahepatic splenic tissue. After tissue biopsy, the definitive diagnosis of low-grade B cell lymphoma on top of intrahepatic splenosis was made.

To our knowledge, this is the first reported case of lymphoma development on top of intrahepatic splenosis.

Ethical approval

This article does not contain any studies with human subjects or animals performed by the authors.

Patient consent

The patient signed a universal consent agreement upon admission to the hospital. This agreement can be provided upon request.

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