Case Report

A case of solitary necrotic nodule of the liver

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Rare association of solitary necrotic nodule of the liver with rheumatoid arthritis and systemic lupus erythematosus

Nil Kocanali[1], Fevzi Duhan Berkan Kilickan[1], Hale Kirimlioglu[2], Murat Gonenc[3], Nurdan Tozun[4,5], Alp Dincer[6,7], Mehmet Erdem Yildiz[7]

[1]Acibadem Mehmet Ali Aydinlar University School of Medicine, Istanbul, Turkiye; [2]Department of Pathology, Acibadem Mehmet Ali Aydinlar University School of Medicine, Istanbul, Turkiye; [3]Department of General Surgery, Acibadem Altunizade Hospital, Istanbul, Turkiye; [4]Department of Gastroenterology, Acibadem Mehmet Ali Aydinlar University School of Medicine, Istanbul, Turkiye; [5]Department of Gastroenterology, Acibadem Altunizade Hospital, Istanbul, Turkiye; [6]Department of Radiology, Acibadem Mehmet Ali Aydinlar University School of Medicine, Istanbul, Turkiye; [7]Department of Radiology, Acibadem Altunizade Hospital, Istanbul, Turkiye

Abstract

Solitary necrotic nodule of the liver (SNNL) is a rare benign lesion with uncertain etiology characterized by a "completely necrotic core" and a hyalinized capsule containing elastin fibers (Journal of Clinical Pathology 36:1181–1183, 1983). We report herein a 26-year-old woman with a previous diagnosis of rheumatoid arthritis, systemic lupus erythematosus, and Sjögren's syndrome and no history of malignancy who presented with a complaint of diarrhea of 1-year duration. In the abdominal ultrasound, multiple paraaortic, portocaval, and ileal lymphadenopathies (LAPs) have been found with the largest one being 2 cm in size. The biopsy of the iliac LAP showed reactive nodular hyperplasia. An abdominal CT disclosed an incidental hypoechoic, heterogenous mass sized 27 × 27 mm close to segment VI of the liver. A trucut biopsy of this lesion was made, and clinicopathologic features of the specimen were compatible with a solitary necrotic nodule of the liver. Here, we discuss the diagnosis and the clinical course of this rare entity in light of current literature.

Keywords: Solitary necrotic nodule of the liver; rheumatoid arthritis; systemic lupus erythematosus.

Introduction

Solitary necrotic nodule of the liver (SNNL) is a rare entity with uncertain etiology. It was first described by Shepherd and Lee[1] in 1983 as an unusual lesion with a "completely necrotic core" and a hyalinized capsule containing elastin fiber but no evidence of malignant tumor in the specimen. The etiology of SNNL is still unknown.[2] The lesion is usually solitary, but multiple necrotic nodules were also reported. Solitary necrotic nodules tend to be in the subcapsular area rather than the deeper regions of the liver.[3] Most of the patients are diagnosed accidentally during investigation for other problems. The association of SNNL with RA and/or other connective tissue diseases is a rarity. We report herein a 26-year-old woman with RA and systemic lupus erythematosus (SLE) and no history of malignancy diagnosed with SLLN at the histopathologic examination of the resected lesion.

Case Report

A 26-year-old woman was admitted to the hospital for the evaluation of diarrhea of 1-year duration.

Her past record revealed that she was diagnosed with rheumatoid arthritis when she was 17 years old. She had complaints of arthralgia, morning stiffness for more than 1 h, and arthritis in the proximal interphalangeal joints of both her hands. Her laboratory tests were rheumatoid factor (+) (242 IU/mL), anti-CCP (+): 1/80, erythrocyte sedimentation rate: 30 mm/h, and antinuclear antibodies (+): 1/200 at the time of diagnosis. C3 and C4 values were normal. She was prescribed methotrexate, hydroxychloroquine, and prednisolone, which she regularly used for 1 year and for another year on demand. She stopped her medications at her will. Five years ago, prednisolone and methotrexate were reinitiated after a flare triggered by a parvovirus infection. She has used her drugs properly since then and was under regular follow-up every 6 months.

Two years ago, she was diagnosed with SLE and Sjögren's syndrome with relevant clinical symptoms (blurred vision and dryness in her eyes, morning stiffness >1 h, and tenderness on multiple joints) and positive anti-SSA and anti-Ro52. She was followed up with no medication, and she is asymptomatic at present. The patient does not use alcohol or tobacco. She reports no history of contact with stray dogs or cats. In her family history, her sister also had seropositive rheumatoid arthritis. Apart from slight abdominal tenderness at the right hypochondrium, a physical examination revealed a normal condition. Blood count, liver aminotransferases, and other biochemical tests were all within normal limits. Stool microscopy and examination for ova/parasites and tests for celiac disease or other causes of diarrhea were normal.

Gastroscopy, colonoscopy, and random biopsies taken from the colon and upper GI tract yielded normal results. Abdominal ultrasound (US) performed 3 months ago showed multiple paraaortic, portocaval, and ileal lymphadenopathies (LAPs), the size of the largest being 2 cm. An abdominal CT disclosed a heterogenous, lobular, slightly hypoechoic mass of size 27 × 27 mm located in the subcapsular region of the posterior segment of the right lobe close to segment VI of the liver (Fig. 1).

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Corresponding author: Nil Kocanali; Acibadem Mehmet Ali Aydinlar Universitesi Tip Fakultesi, Istanbul, Turkiye
Phone: +90 216 500 44 44; **e-mail:** nilkocanali@gmail.com

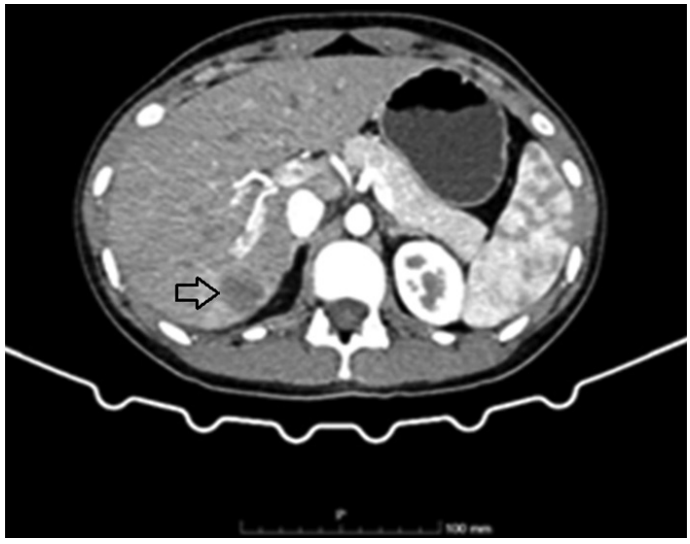

Figure 1. Abdominal CT showing slightly hypoechoic heterogenous solid mass in the subcapsular region of the posterior segment of the right lobe of the liver (see arrow).

Abdominal magnetic resonance imaging (MRI) with contrast confirmed the presence of a round lesion with a diameter of 25.6 mm on its short axis, with a necrotic center and straight, thick contours located at the posterior of segment VI of the liver (Fig. 2a). There was a low signal in T1-weighted images in the center and a high signal in the periphery of the lesion in T2-weighted images. The lesion did not opacify after the contrast injection. In the arterial phase, the pattern of enhancement was heterogenous at the periphery. In the diffusion-weighted study, the lesion showed diffusion restriction in the periphery and lengthening of diffusion in the middle section. There were also many lymphadenopathies over 2 cm located around interaortacaval, hilum of liver, celiac trunk, paraaortic, external and internal iliac areas. The patient underwent surgery for the removal of one of the lymph nodes. The pathology results of the lymphadenopathy were obtained as reactive lymph hyperplasia at histopathology.

After 3 months, a dynamic contrast-enhanced MRI revealed an increase in the diameter of the lesion up to 31.0 mm with regression of paraaortic and interaortacaval lympadenopathies (Fig. 2b).

The patient was seronegative for hepatitis B and hepatitis C virus markers. The QuantiFERON test was negative, and alpha-fetoprotein was within normal limits. As the radiological findings of the lesion in the liver were not compatible with any benign tumor and the size of the mass had increased within a few months, a US-guided trucut biopsy was performed. The histopathologic examination defined the liver lesion as chronic granulomatous inflammation and necrosis surrounded with fibrotic tissue, with a probable diagnosis of the solitary necrotic nodule. There was no evidence of malignancy. Sclerosing hemangioma, inflammatory pseudotumor, and inflammatory myofibroblastic tumor of the liver were considered in the differential diagnosis. Immunostains for smooth muscle actin, CD68 showed focal positivity at the inflammatory areas, CD34 and desmin were negative. Histochemistry stain periodic acid–Schiff (PAS) stain was negative. No fungal hyphae or spores and no parasites were observed.

With these findings, the patient underwent surgery and the nodule close to segment VI was completely removed.

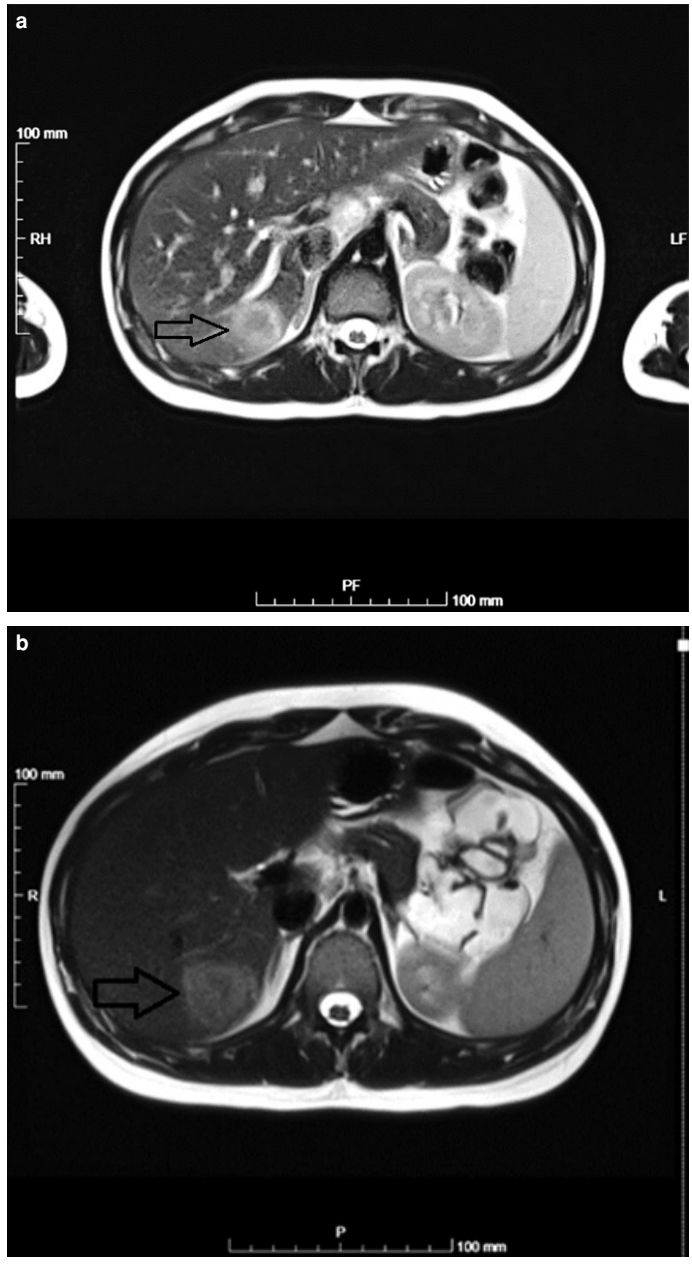

Figure 2. (a) Abdominal MRI T2 showing a mass hypointense in the center and hyperintense in the periphery at segment VI of the liver. **(b)** Dynamic contrast MRI enterography showing the progression of this lesion 3 months after the first abdominal MRI (see arrow).

At the histologic examination of the mass, a localized necrotizing granulomatous lesion suggestive of a rheumatoid nodule was observed (Fig. 3a). Masson's trichrome stain demonstrated the nodule consisted of fibrous tissue surrounding a center of fibrinoid necrosis and the focal hyalinizing fibrotic areas of the central necrotic area (Fig. 3b). There was a surrounding palisade of histiocytes, fibroblasts, and mixed infiltrate of lymphocytes, plasma cells and occasional eosinophils and neutrophils (Fig. 3c).

One year after the resection of the liver nodule, the patient was in full health, and her diarrhea improved after appropriate treatment for IBS.

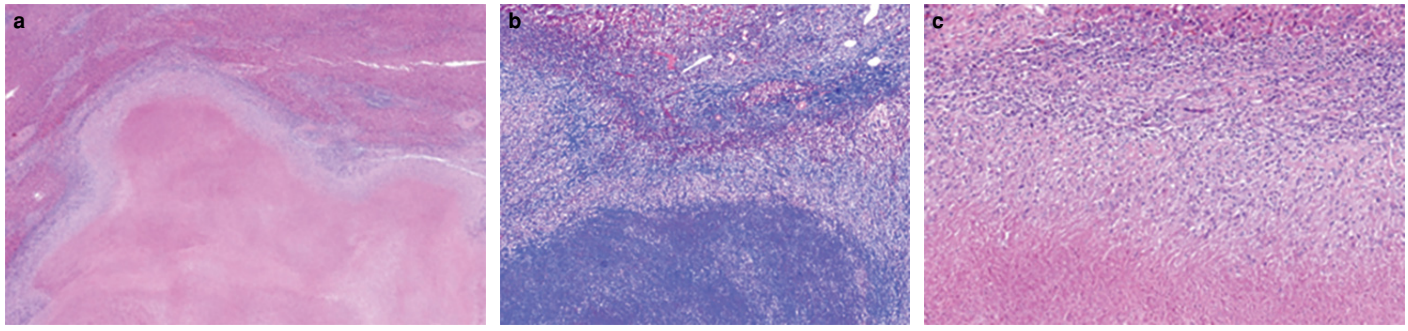

Figure 3. (a) ocalized necrotizing granulomatous lesion (HE 100×). **(b)** Fibrous tissue surrounding a center of fibrinoid necrosis and hyalinizing fibrotic areas of the central necrotic area (Masson's trichrome 200×). **(c)** Surrounding granulomatous inflammation (HE 400×).

Discussion

SNNL is a rare lesion that is characterized by a nodule with a necrotic core surrounded by a hyalinized fibrotic capsule with elastic fibers.[1] Most cases are accidentally found in patients with normal liver function.[4] Reported cases are usually males in the seventh or eighth decade, but as in our case, young patients have also been reported. The entity has been linked to sclerosing hemangioma, trauma, necrotic parasitic infections, hepatic infarctions, organized thrombi, benign lesions, or distant effects of intra-abdominal tumors. Remnants of parasitic organisms and hemangiomas have been shown histologically in some studies.[5,6] De Luca et al. favor the ischemic hypothesis, which was mentioned in a few cases with partial stenosis in the celiac trunk. The lesion may remain stable for years, but there are reports of enlargement or reduction in size in the literature.[7,8] In our case, an increase in the size of the nodule of 8 mm within 3 months prompted us to undertake surgical resection.

In the differential diagnosis of SNNL, benign liver lesions, metastatic tumors, rheumatoid nodules, sarcoidosis, infections, and even tuberculomas must be considered clinically, radiologically, and histologically.[2] Abdominal imaging often leads to misinterpretation of this lesion as metastasis.

On the other hand, the liver is commonly involved in connective tissue diseases. Rheumatoid arthritis may be associated with nodular regenerative hyperplasia, and rarely, rheumatoid nodules may occur in solid organs, such as the lungs and heart. However, their presence in the liver is an exceptional finding.[9,10]

The association of SNNL with SLE is rare. In our case, both the RA and LE were considered probable causes of SNNL.

Although SNNLs are considered benign, the uncertainty of the radiological findings and the difficulty of differential diagnosis from malignant lesions may lead the clinician to do a liver biopsy. Occasionally, surgical resection may be necessary for the confirmation of the diagnosis, especially when imaging studies show a progression of the lesion under follow-up, as was the case in our patient.

In conclusion, SNNLs are rare entities diagnosed accidentally during a check-up. The association of SNNL with RA and/or other connective tissue diseases is rare. Our 26-year-old female patient's SNNL was associated with RA and SLE although the presence of this lesion has been linked to trauma, parasitic infections, hepatic infarctions, and thrombi, benign lesions, or distant effects of intra-abdominal tumors in the literature.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Conflict of Interest: The authors have no conflict of interest to declare.

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