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Isolated Hepatic Sarcoidosis Mimicking Liver Metastasis: A Case Report

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Abstract

Sarcoidosis is a multisystemic, non-caseous granulomatous disease, of unknown etiology which mainly affects the lungs and mediastinal lymph nodes. Liver involvement is commonly seen in multisystemic sarcoidosis, yet; isolated primary hepatic sarcoid localization is rare, and can be misleading as it may mimic malignant lesions. We describe a rare case of a-27-years old female, who presented with chronic abdominal pain. Abdominal imaging showed nodular hepatic lesions suggestive of metastasis, however; liver biopsy revealed evidence of non-caseating epithelioid cell granulomas with Langhans multinucleated giant cells suggesting hepatic sarcoidosis.

Keywords: *Liver sarcoidosis; Liver metastasis; Abdominal pain*

Case Presentation

We report the case of a 27-year-old female who presented to the gastroenterology department of our hospital with episodic, long-standing abdominal pain localized to the right upper quadrant. The patient denied intestinal transit disorders and weight loss. No particular medical or surgical history was found. Physical examination showed no anomalies. Routine laboratory test parameters were within normal limits. Abdominal ultrasound was initially performed. It revealed hepatomegaly with multiples rounded, well defined hypoechoic nodular lesions. The patient underwent an abdominal MRI for a better characterization. It revealed an enlarged liver containing multiple hyperintense nodules with central hypo signal on T2 weighted and T2-weighted fat-saturated images. They demonstrated a low signal on T1 weighted images, as compared with the adjacent liver parenchyma. Diffusion-weighted sequences revealed nodular high signal intensity. On post contrast T1-weighted fat-saturated images, hepatic nodules described delayed, heterogenous enhancement and intrahepatic vascular and biliary structures adjacent to the lesions seemed to be intact. Given the possibility of metastatic origin, a thoraco-abdominopelvic contrast enhanced CT was performed, but showed no potentially malignant lesion aside of hypoattenuating hepatic nodular lesions that enhanced on post contrast images. Ultrasound-guided liver biopsy was performed. Histological specimen showed evidence of multinucleated giant cells together with non-caseating epithelioid cell granulomas in the portal and periportal spaces. Hepatic sarcoidosis was diagnosed based on such pathological findings. The patient was then treated with corticosteroid therapy and showed good response (Figures 1-4).

Discussion

Sarcoidosis is an idiopathic immune-mediated systemic disease, characterized by the presence of non-caseating epithelioid granulomas in the absence of other causes of granulomatous reactions [1,2]. Sarcoidosis prevalence is highest in African Americans and Scandinavian ethnicities. Its main peak of incidence is described for the age group 20–39 years for both genders, with women having a second incidence peak at the ages of 65–69 years [1-3]. Lungs and mediastinal lymph nodes are the mainly targeted sites. Concomitant extrapulmonary is described in 30% of cases. Commonly involved localizations are the

abdominal cavity, the skin, the eyes, the central nervous system and the heart. Isolated extra pulmonary involvement remains a rare entity with only 2% of reported sarcoidosis cases [4]. On the other hand, liver involvement was found in 50-80% in patients with multisystemic sarcoidosis; however, isolated hepatic sarcoidosis remains rare; it has been reported only in 13% of cases [5].

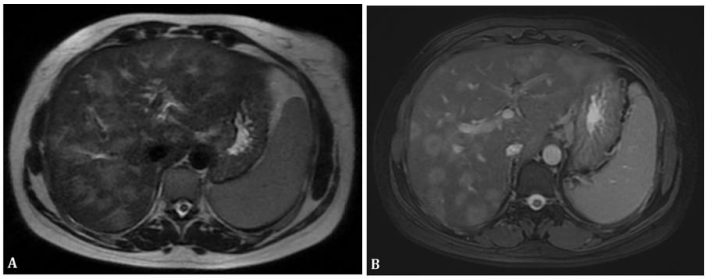

Figure 1: *Axial T2 weighted (A) and T2 fat-saturated (B) abdominal MRI images demonstrating enlarged liver with multiple hyperintense nodular lesions.*

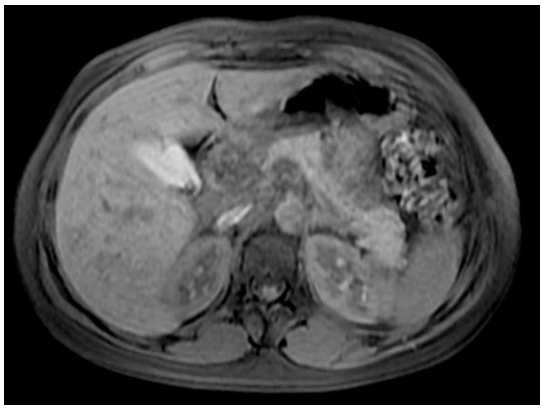

Figure 2: *Axial T1 weighted abdominal MRI image demonstrating multiple hypointense hepatic nodular lesions.*

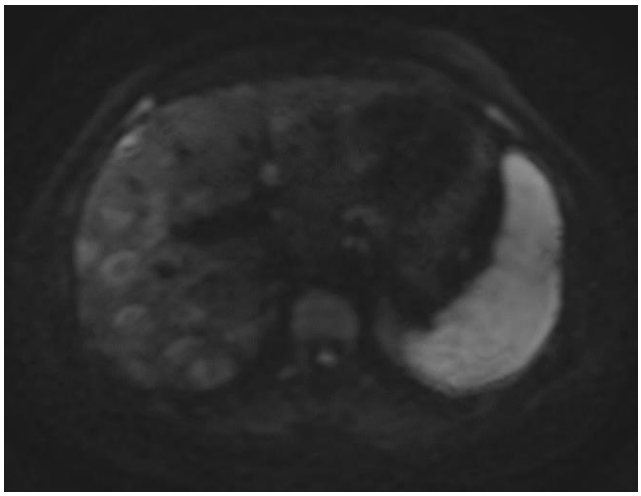

Figure 3: *Axial diffusion weighted (DWI) abdominal MRI image demonstrating hyperintense hepatic nodular lesions.*

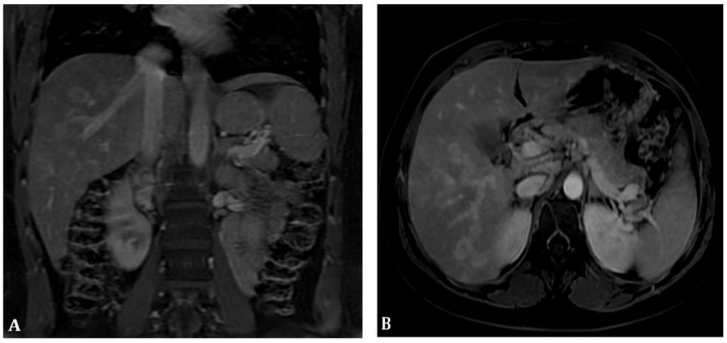

Figure 4: *Coronal (A) and axial (B) T1 weighted Fat-saturated post contrast abdominal MRI images demonstrating enhancing hepatic nodular lesions.*

Hepatic sarcoidosis can present with a board spectrum of manifestations which varies from asymptomatic to non-specific signs such as abdominal pain, nausea, vomiting, fever, arthralgias and right hypochondrium tenderness. Hepatosplenomegaly is described in 10-40% of cases. Liver cirrhosis and portal hypertension are mostly seen in long standing cases [6]. Liver function tests are abnormal in 2-60% of cases. Alkaline phosphatase level is most commonly affected [5]. Other blood tests are non-specific, however elevated serum angiotensin converting enzyme (s-ACE) may help narrow differential diagnosis [1]. Imaging modalities play an important role in assessing the diagnosis and follow up of patients with extrapulmonary sarcoidosis. Abdominal ultrasonography may demonstrate hepatomegaly with increased liver echogenicity, which can be either homogenous or diffusely heterogeneous. Focal nodules, when visible, are round or oval shaped, hypoechoic, diffusely distributed, ranging in size of 1-2mm to centimeters, with a tendency of confluence, hypo vascular on Color Doppler and localized in the portal and periportal spaces. Associated abdominal adenopathies are found in 76% of cases. Enlarged lymph nodes are usually periportal, celiac, paracaval and para-aortic, with dimensions ranging from 1 cm to 6 cm [6]. Computed tomography (CT) may reveal hepatomegaly with possible low-density intrahepatic septa. Liver nodules are hypodense with no mass effect nor peripheral enhancement. CT scan is also useful in the diagnosis of liver cirrhosis and portal hypertension subsequent to sarcoidosis [3]. Focal calcifications are uncommon, and usually presents as round or oval hyperdense, homogeneous foci on non-enhanced images [7]. MRI typically demonstrates hypointense and hypoenhancing nodules relative to the adjacent liver parenchyma, with no impact on the surrounding parenchyma or adjacent vessels. T2-weighted fat-saturated images are the most conclusive sequences that help differentiate sarcoid nodules from malignancies, as these appear most frequently hyperintense [3]. However, signal intensity depicted on T2-weighted images varies according to the degree of activity of

the disease: in case of active inflammation, nodules are hyperintense with restriction on diffusion weighted imaging (DWI), whereas, fibrotic nodules show low signal on T2-weighted and diffusion sequences [4]. In our case, hepatic nodules demonstrated high signal intensity on T2 weighted and diffusion images with slightly homogenous enhancement on post contrast sequences, which suggests high inflammation activity. Moreover, absence of mass effect and intact vascular and biliary structure is highly suggestive of benign or infiltrative disease [8]. Other signs suggestive of sarcoidosis are irregular contour of the liver and high periportal signal intensity. MR cholangiopancreatography is useful in assessing biliary stenosis and dilatation subsequent to compressive hilar nodules and lymph nodes [3]. CT and MRI are useful not only for diagnostic purposes but also for follow-up of hepatic lesions, and to verify the treatment efficiency. They represent a valid diagnostic aid that can assess the diagnosis of sarcoidosis in many cases, especially in the presence of characteristic clinical features, however, definitive diagnosis is based on the biopsy with the demonstration of non-caseating epithelioid granulomas [1].

The differential diagnosis of isolated liver nodules includes primitive benign and malignant tumors, metastasis, tuberculosis, lymphoma, liver abscesses and vascular lesions. Treatment is based on the symptoms and severity of the disease. Asymptomatic patients require no treatment; whereas, in symptomatic cases, corticosteroids and/or ursodeoxcholic acid may be considered. Other immunosuppressive agents may be required in case of non-response to first line treatment [1].

Conclusion

Liver sarcoidosis is a benign entity that can mimic liver metastasis. The radiological presentation may vary but some subtle signs such as the absence of masse effect on adjacent structure can help assess the diagnosis. Long stranding sarcoidosis can progress to end-stage liver disease; hence it should always be considered as a differential diagnosis so it can be clearly diagnosed and carefully monitored.

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