

Multiple Facets of Abdominal Sarcoidosis

Keywords: Sarcoidosis; Liver; Spleen; Ultrasound; Computed tomography; Magnetic resonance imaging

Abbreviations: US: Ultrasound; CECT: Contrast Enhancement Computed Tomography; MRI: Magnetic Resonance Imaging; CEUS: Contrast Enhanced Ultrasound

Editorial

The complexity of sarcoidosis is in part due to its multiple clinical manifestations that not only involve organs such as lungs but also sites such as liver and spleen. Abdominal manifestations of sarcoidosis are common if paucy- or asymptomatic forms of the disease are considered, with a reported frequency of 50-70% [1]. Virtually every organ can be involved by sarcoid lesions, but liver and spleen are the sites most often affected. Rarely pancreas, gastrointestinal and urinary system have been described as organs involved from sarcoidosis [2-5].

Increase of liver enzymes (e.g., transaminases, gamma-gt and alkaline phosphatases) is commonly observed in patients with sarcoidosis [6], but should not be considered a priori evidence of hepatic involvement in patients affected by pulmonary form of sarcoidosis unless confirmed by biopsy or, at least, by imaging. Other concomitant etiologies can indeed occur such as non-alcoholic fatty liver disease and viral hepatitis and differential diagnosis can be challenging if a high clinical suspect does not occur in these situations [7,8].

Besides asymptomatic form, sarcoidosis of the liver can manifest clinically with a wide range of symptoms, such as anorexia, weight loss, night sweats and fever. Symptoms derived from intrahepatic cholestasis (e.g., jaundice and pruritus), liver failure or those derived from portal hypertension (e.g., ascites, bleeding from gastroesophageal varices rupture) due to granulomas involving the portal triad can be rarely observed. Sometimes, peritoneum can be infiltrated by multiple nodules mimicking peritoneal carcinomatosis [9].

If clinically suspected, abdominal involvement by sarcoidosis can be first detected by imaging studies. Conventional ultrasound (US), followed by second imaging techniques such as contrast enhancement computed tomography (CECT) or magnetic resonance imaging (MRI) shows homogenous hepatomegaly, fatty liver disease-like pattern with increase echogenicity, parenchymal inhomogeneity with coarsening pattern or multiple prominent nodules. Rarely, isolated hepatic nodules can be found. Such masses should be differentiated with focal malignant lesions. Other common findings are splenomegaly, with or without prominent nodules and lymph node enlargement [7,9-11]. Overcoming common limits of traditional contrast-imaging techniques, contrast-enhanced ultrasound (CEUS), a novel safe US technique, has documented reliability to detect nodules in some case series, where conventional US was usually negative [8,12,13].

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Editorial

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Histopathologically, granulomas are characteristically non-caseating and characterized by accumulation of histiocytes, lymphocytes, sparse lymphoid and a variable amount of fibrosis and collagen. The term granuloma derives from the Latin granum (grain) and means the typical gross appearance of these lesions [14]. These lesions seem to represent a continuous inflammation process due to incomplete antigen degradation associated with chronic activity of cell mediated immunity [15]. The most important problem of abdominal sarcoidosis, but also of systemic disease, is the wide range of disorders that comes into differential diagnosis. Sarcoidosis can manifest with similar clinical and radiological findings of conditions such as tuberculosis, lymphoma, and neoplastic disorders. It should not surprise therefore that definitive diagnosis is very difficult to obtain in some occasions, and these considerations are even more true if both conditions (e.g., sarcoidosis and malignant conditions) are associated in the same patient. Several studies have demonstrated that sarcoidosis can be a risk factor for malignant degeneration, where continuous inflammatory process represents the leading mechanism [15-17]. A correct diagnosis is therefore essential and biopsy is necessary to demonstrate non-caseating granulomas. Sites for biopsy can be liver and visceral lymph nodes if more accessible sites (e.g., skin, superficial lymph nodes) are not available. Spleen has been considered in the past as an additional site of biopsy [18] but can increase the risk of serious complications (e.g., bleeding, GI perforation). In conclusion, a definitive diagnosis of abdominal sarcoidosis should be globally based on a thorough clinical algorithm with evaluation of the patient, laboratory tests, biopsy from accessible site that demonstrates characteristic granulomatous lesions and on the exclusion of infectious and/or non-infectious immunogenic factors from the epithelioid cell granulomas [19-22].

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