Intra-abdominal sarcoidosis: radiologic manifestations

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Learning objectives

Intra-abdominal involvement of sarcoidosis can be found in the absence of pulmonary or lymphatic disease (1). Liver (50-80%), spleen (40-80%), lymph nodes (30%) and kidney are more commonly involved, sometimes without symptoms. The lesions are less characteristic, mimicking more common neoplastic or infectious diseases such as lymphoma, diffuse metastasis, granulomatous or mycobacterial infections (2). In this educational exhibit we review the imaging manifestations of intra-abdominal sarcoidosis.

Background

Sarcoidosis is a multisystem inflammatory disease of unknown aetiology, characterized by non-caseating epithelioid cell granulomas in the absence of other granulomatous diseases such as tuberculosis, fungal infections, autoimmune processes, or delayed-type hypersensitivity to foreign antigens (3).

Epidemiological reports of sarcoidosis vary among racial groups, but the worldwide prevalence is between 2-60 per 100,000 people (4). The disease is usually diagnosed between 20-40 years of age (5). Women are more frequently affected than men (5). It is believed that the disease arises in genetically susceptible individuals who are exposed to deleterious environmental agents including viruses such as herpes viruses, retroviruses, and cytomegalovirus, as well as bacteria including Borrelia burgdorferi, mycobacteria and mycoplasma.

Diagnosis of sarcoidosis is generally based upon clinical features, demonstration of non-caseating granulomas in at least two different organs, negative staining and culture for acid fast bacilli, absence of exposure to toxins, and lack of drug-induced disease.

The lungs (> 90%) and the lymphoid system (30%) are involved most commonly (1). Extrapulmonary involvement of sarcoidosis is reported in 50% of cases and the abdomen is the most common site with a frequency of 50%-70%. Although it is usually asymptomatic, its presence may affect the prognosis and treatment options.

Corticosteroids should be instituted when organ function is threatened. The overall prognosis is good although most patients will have some permanent organ impairment. Cardiac and pulmonary involvements are the main causes of death.
Findings and procedure details

Sarcoidosis of the Liver

Liver follows lymph nodes and lung in the frequency of involvement. Furthermore, it is reported that a significant fraction (26%) have liver lesions without lung involvement (6). About 50-79% of livers are involved by biopsy and 67-70% by autopsy (7, 8).

The commonest radiologic manifestation of hepatic sarcoidosis is hepatomegaly, which is found in about more than half of the patients on abdominal computed tomography (CT) scans (9). It is often together with splenomegaly. Other findings range from asymptomatic incidental granulomas to portal hypertension and cirrhosis from granulomas in the portal triad and fibrosis due to chronic inflammation. Portal vein thrombosis is reported as a frequent complication of hepatic sarcoidosis possibly as a result of stasis from obliteration of small portal veins (10). It is also suggested that there is an additional correlation between chronic hepatic sarcoidosis and hepatocellular carcinoma (11).

Ultrasound (US) findings include parenchymal echogenicity, coarsening of the liver parenchyma with or without discrete nodules and contour irregularity (12). Calcification is uncommon but can be seen with long-standing disease. The nodules which represent the coalescence of small granulomas are typically innumerable, diffusely distributed and range from 1 to 2 mm to several centimeters in size. They have been reported to be hypoechoic on US and hypodense on computed tomography (CT) scans, relative to the background liver parenchyma (13). However, they may also be hyperechoic based on the background liver echogenicity and degree of fibrosis present in the granuloma. Focal nodules are the lesions identified in 5% of patients at imaging (14). On MRI, the lesions are hypointense on all sequences most conspicuous on the T2-weighted fat-saturated images (15). They enhance less than the background liver on gadolinium-enhanced T1-weighted images (15). Rarely, an increased signal intensity of diffusely involved liver is detected on T2-weighted images when compared with the normal spleen. Figure 1a, 1c, 1d, and Figure 2 shows US, CT and MR images of granulomatous lesions in hepatic sarcoidosis.

Sarcoidosis of the Spleen

By fine needle aspiration and angiography, the frequency of splenic involvement has been reported in 24-53% of cases (16). Splenomegaly from sarcoidosis is usually associated with multiple organ involvement, commonly the lungs and the liver. Splenic infiltration can be homogeneous or in the form of multiple nodules (17). Splenic nodules can be seen in 15% of the cases undergoing CT scanning of the abdomen and are more common than the hepatic ones (14). Similar to the hepatic nodules, the splenic nodules
are seen as multiple, hypodense, hypointense and non-enhancing lesions, scattered in the spleen. They tend to be discrete but may coalesce with increasing size (15). The differential diagnosis of low-attenuation nodules in the liver and spleen includes infection, metastatic disease, and lymphoma. Figure 1b, 1c, and 1d shows US, CT and MR images of granulomatous lesions in splenic sarcoidosis.

**Sarcoidosis of the Gastrointestinal System**

Sarcoidosis of the gastrointestinal (GI) tract is extremely rare and the majority of the cases are asymptomatic. On autopsy, approximately 10% of systemic sarcoidosis patients were found to have gastric granulomatous mucosal infiltration, most often in the antrum (18). Antral narrowing and deformity which leads to gastric outlet obstruction may develop due to gastric mucosal fold enlargement mimicking Menetrier's disease (19, 20). A segmental linitis plastica-type picture which should be differentiated from gastric carcinoma is the most common abnormality found on upper GI series (19). Extrinsic compression from extensive retroperitoneal lymphadenopathy may also occur throughout the GI tract.

Small bowel involvement is the least common site of all in gut sarcoidosis (21). Extrinsic compression by abdominal and mesenteric lymph nodes or reduction of the lumen size secondary to cicatrizng constriction in consequence of mucosal granulomatous lesions, results in intestinal obstruction (1, 22).

Colorectal sarcoidosis can occur in the presence of grossly normal appearing mucosa. Colonic sarcoidosis involves multiple nodules, polyps, stenosis, obstructive lesions, aphthous erosions, or ulcers (23, 24, 25). Figure 3c shows barium small bowel enema in a patient with terminal ileum and ileocecal involvement in a patient with sarcoidosis.

**Sarcoidosis of the Peritoneum**

Sarcoidosis rarely affects the peritoneum (26). The most frequent clinical presentation is exudative ascites, multiple granulomatous nodules studding the peritoneum or a single peritoneal lesion. Peritoneal biopsy is required due to close gross visual resemblance, to rule out carcinomatosis, tuberculosis and fungal infections (1, 22). Figure 3a, 3b, and 3d shows CT images of nodal, omental and mesenteric involvement in a patient with sarcoidosis.
Enlarged lymph nodes are detected in approximately 30% of patients particularly in the porta hepatis, para-aortic region, and celiac axis (14). Lymph nodes are typically smaller (less than 2 cm in diameter) and more discrete (rather than confluent) and less commonly involve the retrocrural area in sarcoidosis than lymphoma (27). Figure 1a shows periportal hypoechoic enlarged lymph node in a patient with sarcoidosis.

**Sarcoidosis of the Pancreas**

Pancreatic sarcoidosis has been detected on autopsy in 1-3% of cases with systemic disease but it is rarely symptomatic during life (28). Typical symptoms of pancreatic involvement are due to direct tissue infiltration, duct obstruction, or constrictive peripancreatic lymphadenopathy.

**Sarcoidosis of the Biliary System and Gall Bladder**

Extrinsic compression of the cystic duct by granulomatous lymph nodes or granulomatous inflammation of the gallbladder may lead acute cholecystitis as a complication of sarcoidosis (29). Subacute or chronic cholecystitis with granulomas in the gallbladder wall has been reported as a complication of sarcoidosis (30).

**Sarcoidosis of the Kidney**

Nephrocalcinosis, nephrolithiasis, and interstitial calcium deposition have been reported in sarcoidosis and may lead to renal failure. Interstitial nephritis is a possible manifestation, and may demonstrate a striated nephrogram on contrast-enhanced CT (31). Glomerular nephritis from direct infiltration may also occur (32).

Nephrocalcinosis is the most frequent effect of sarcoidosis on the kidney. It is in consequence of hypercalcemia or hypercalciuria secondary to granulomatous production of calcitriol (32). These granulomata are typically extrarenal (33). Direct granulomatous involvement of the kidneys is rarely observed and is indistinguishable from lymphoma or metastasis (34).

**Sarcoidosis of the Genital Tract**

The most common site of involvement of the female reproductive system is the uterus (35). It is usually detected during the investigation of abnormal uterine bleeding in patients with a previous history of sarcoidosis at other anatomic sites (36-39). Ovarian sarcoidosis
is extremely rare and known to mimic ovarian malignancy with soft tissue nodules (35). There are no specific radiological findings in the literature to describe ovarian sarcoidosis (35).

Sarcoidosis involving the male reproductive tract has rarely been reported. The epididymis, testis, and prostate gland, with only rare involvement of the spermatic cord, scrotum, and penis are reported in order of decreasing frequency (40). The lesions exhibit enhancement on gadolinium-based intravenous contrast administration T1-weighted images and low signal intensity on T2-weighted images (41).

Images for this section:

**Fig. 1:** a. Axial ultrasound image shows multiple, diffusely distributed, small, hypoechoic, rounded granulomas in enlarged liver and periportal hypoechoic enlarged lymph node (arrow). b. Sagittal ultrasound image shows 3.5 cm sized, hypoechoic, focal splenic lesion. c. Axial contrast enhanced CT image shows contour irregularity of the enlarged liver. Multiple, diffusely distributed hypodense granulomas ranging from 1 to 2 cm are detected in the liver and spleen. d. Axial contrast enhanced CT image shows irregular,
large, hypodense areas representing coalescence of the granulomas in the liver and hypodense granulomas ranging from 1 to 2 cm in the spleen.

Fig. 2: a. Axial ultrasound image shows 1 cm sized, hypoechoic, rounded granuloma in the peripheral subcapsular region of the liver. b. Axial T1W-GRE image demonstrates hypointensity of the same lesion.
Fig. 3: a. Axial contrast enhanced CT image shows pericecal multiple enlarged lymph nodes. b. Subsequent CT examination demonstrates wall thickening of the ileocecal area, heterogeneous hyperdensity of the omentum and also the mesenteric free fluid. c. Barium small bowel enema shows mucosal irregularity, nodularity and angulation of terminal ileum and ileocecal area. d. Coronal reformatted CT image shows multiple lymph nodes in the mesenteric root of the same region.
Conclusion

Sarcoidosis is a multisystem inflammatory disease which has been reported in every system and organ of the human body. Intra-abdominal sarcoidosis is less common, but most often asymptomatic. Long-standing unrecognized disease can result in significant morbidity and mortality. It may mimic more common infectious or neoplastic conditions. Sarcoidosis should be included in the differential diagnosis to avoid complications. Imaging contributes diagnosis and management of intra-abdominal sarcoidosis.

Personal information

References


