

Appendiceal sarcoidosis presenting as acute appendicitis

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ABSTRACT

Sarcoidosis is a chronic inflammatory multisystem disease. The stomach is the most commonly involved gastrointestinal organ. Symptomatic appendicular sarcoidosis is extremely rare. We present a case of a 49-year-old woman with abdominal pain. An ultrasound of the abdomen was suggestive of acute appendicitis. Laparoscopic appendectomy was performed and the pathology revealed nonnecrotizing granulomas. Biopsy of the mediastinal lymph nodes suggested noncaseating granulomas. She was treated with steroid therapy followed by mycophenolate mofetil. Our case demonstrates the importance of considering appendiceal sarcoid among the differentials in a patient with systemic sarcoidosis presenting with an acute abdomen.

KEYWORDS Acute appendicitis; appendiceal neoplasm; appendiceal sarcoidosis; gastrointestinal sarcoidosis

Sarcoidosis is a chronic inflammatory multisystem disease.¹ Organs typically affected include the lungs, lymph nodes, and skin.² Gastrointestinal (GI) involvement, although not uncommon, is asymptomatic 99% of the time.³ The stomach is the most commonly involved GI organ, but the esophagus, appendix, colon, and rectum have also been implicated.⁴ Fewer than 10 cases of appendiceal sarcoidosis have been reported in the English language literature. Symptomatic appendiceal sarcoidosis is extremely rare.

CASE DESCRIPTION

A 49-year-old black woman with recurrent chronic low back pain and kidney stones presented to the emergency department with worsening abdominal pain for 3 days. The pain was sharp, located in the right iliac fossa, and radiated to the periumbilical region.

The patient was hemodynamically stable on arrival with a blood pressure of 120/70 mm Hg and a heart rate of 103 beats/minute. Palpation of the right iliac fossa disclosed tenderness. One hour after admission, the patient had an episode of nonbilious vomiting containing food particles. Her hemoglobin level was 11.2 g/dL; sedimentation rate, 9 mm/h; and white blood cell count, $4.2 \times 10^3/\mu\text{L}$. An ultrasound of the abdomen was suggestive of acute appendicitis with McBurney's point

tenderness. A computed tomography (CT) scan of the abdomen showed an appendicular mass. Magnetic resonance imaging (MRI) of the spine done just prior to this admission (for back pain) showed hypoechoic lesions concerning for vertebral metastases. A fluorodeoxyglucose–positron emission tomography scan done at the same time revealed extensive metastatic lesions with nodular uptake in the right upper lobe of the lung, numerous mediastinal lymph nodes, hypermetabolic foci in the axial skeleton, and focal hypermetabolism in the mid to distal appendix (standardized uptake value 3.8), concerning for an appendiceal neoplasm (*Figure 1*).

Due to concern for perforation of the appendicular mass and abscess formation in the setting of appendicitis, a laparoscopic appendectomy was performed and the histologic study revealed nonnecrotizing granulomas. After surgery, the patient's abdominal pain resolved. A CT scan of the chest was performed to further delineate the mediastinal lymphadenopathy (*Figure 2a*). Subsequently, a bronchoscopic ultrasound-guided biopsy of the lymph nodes revealed granulomas without active inflammation (*Figure 2b*). Serum angiotensin-converting enzyme levels were elevated (68 U/L).

Oral corticosteroids were initiated at 30 mg daily (0.5 mg/kg/day) prior to discharge. Her back pain improved over a few months. A repeat MRI of her spine after 6 months of treatment

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Received February 20, 2020; Revised March 11, 2020; Accepted March 16, 2020.

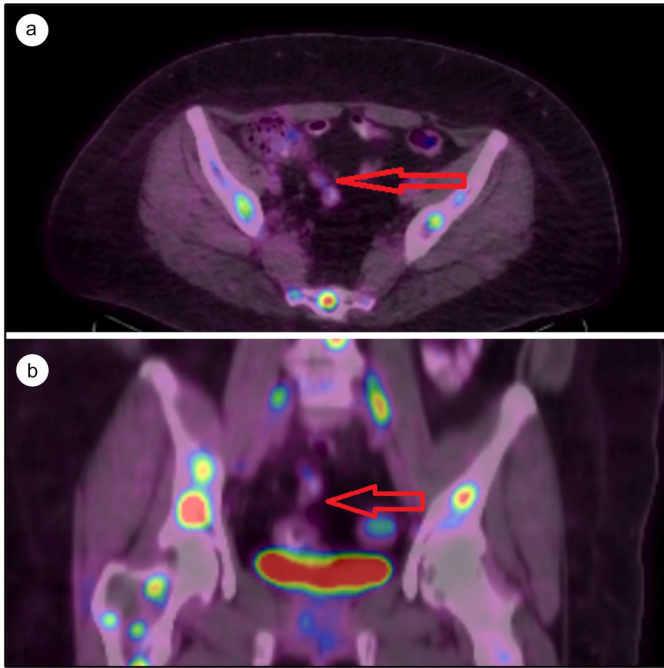


Figure 1. Fluorodeoxyglucose positron emission tomography scan of the abdomen: (a) axial section, with the arrow pointing to the appendiceal mass, and (b) coronal section, with the arrow pointing to the appendiceal mass.

showed a decrease in the size of the vertebral lesions. Steroids were slowly tapered by 5 mg per week. At her 6-month visit, mycophenolate mofetil was initiated at 500 mg twice daily and the dose was increased by 500 mg weekly as tolerated. Currently, she remains symptom free on mycophenolate (1500 mg twice daily) and prednisone (5 mg daily).

DISCUSSION

Fewer than 10 cases of sarcoid involving the appendix have been reported, of which <4 cases reported nonnecrotizing granulomas without acute inflammation.⁵ The present case of appendiceal sarcoidosis causing acute right iliac fossa pain without evidence of acute inflammation appears to be the fifth with similar findings.⁶ In our patient, granulomas identified after appendectomy provided one of the major clues for the diagnosis. Her pain completely resolved postoperatively.

Although there are no specific guidelines for the treatment of GI sarcoidosis, typically systemic sarcoidosis is treated for at least a year and then imaging is repeated to determine the disease status. For most cases of GI sarcoidosis, steroids are the standard immunosuppressants and are usually initiated at 0.5 mg/kg/day. They can be titrated to 10 mg/day over 6 months based on the clinical response. Steroid-sparing agents like methotrexate, azathioprine, leflunomide, and mycophenolate mofetil can be started at low doses and titrated up.^{7–9}

In conclusion, GI sarcoidosis may present with symptoms of acute appendicitis without inflammation. Our case demonstrates the importance of considering appendiceal sarcoid among the differentials in a patient with systemic sarcoidosis

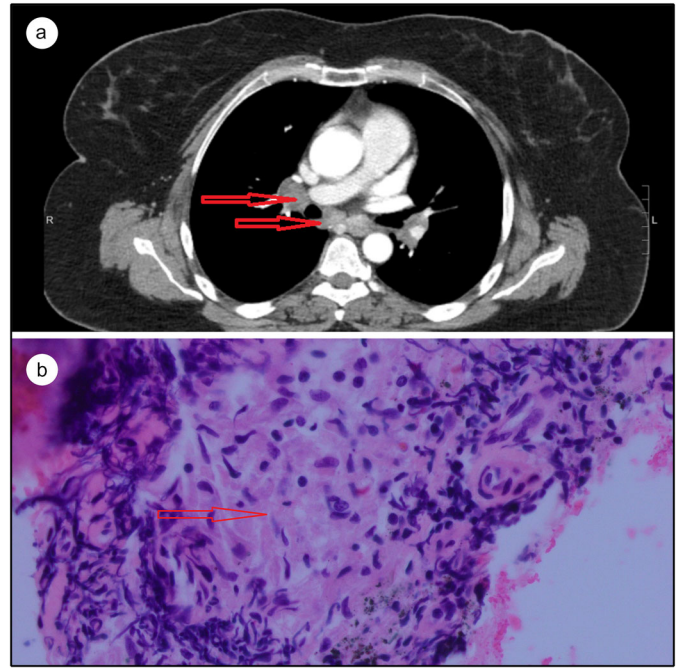


Figure 2. (a) CT of the chest, with the arrows pointing to mediastinal lymphadenopathy. (b) Lymph node biopsy with hematoxylin and eosin stain showing noncaseating granuloma.

presenting with an acute abdomen. It is important to operate on these patients emergently to avoid the risk of perforation.⁶ Steroid therapy and or steroid-sparing immunosuppressants should be considered in the postoperative period.

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