Unusual association of diseases/symptoms

Appendicular sarcoidosis mimicking acute appendicitis

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Summary

Appendicular sarcoidosis is a very rare cause of acute abdominal pain, with only seven cases reported previously in the literature. A 45-year-old woman, known to have sarcoidosis, presented to the emergency department with a 1-week history of epigastric and right iliac fossa abdominal pain. At diagnostic laparoscopy, an acutely inflamed appendix was found and removed as well as an omental mass which was biopsied. Subsequent histopathological examination of the appendix demonstrated appendicular sarcoidosis without acute appendicitis and chronic inflammatory changes in the omental biopsy. The patients' symptoms completely resolved postoperatively. It is important to undertake urgent operative intervention in patients with sarcoidosis who present with right iliac fossa pain, owing to the high risk of perforation.

BACKGROUND

Sarcoidosis is a chronic, inflammatory, granulomatous multisystem disease of unclear aetiology, with a notably higher prevalence in African-American populations. There are two peaks of incidence; 25–35 and 45–65 years. Organs typically affected include the lungs, lymph nodes and skin. Involvement of the gastrointestinal system, although not uncommon, is usually asymptomatic. In patients who are known to have sarcoidosis, operative intervention should not be delayed because of the high risk of perforation.

CASE PRESENTATION

A 45-year-old South Asian lady presented to the emergency department with a 1-week history of worsening epigastric and right upper quadrant pain, with no associated vomiting or melena. Her medical history was significant for sarcoidosis, a perforated duodenal ulcer, a hiatus hernia, early menopause following sterilisation and protein S deficiency. Her regular medications included hydroxychloroquine 200 mg once a day and lansoprazole 15 mg once a day. The patient had previously declined steroid immunosuppression for her sarcoidosis and warfarin for her protein S deficiency.

On examination, she was afebrile but tachycardic with a heart rate of 127 beats/min. She was initially tender in the epigastrium and right upper quadrant, later migrating to the right iliac fossa.

INVESTIGATIONS

The patient's white cell count was 6.1×10^9 cells/l, and C reactive protein was 59.7 mg/l. Her urea and electrolytes and liver function tests were normal, except for a bilirubin of 26 mg/dl. Urinalysis was unremarkable. Chest x-ray revealed bilateral hilar and right para-tracheal lymphadenopathy (figure 1).

Abdominal ultrasound demonstrated free fluid in the pouch of Douglas, extending into the right iliac fossa,

where thickened loops of bowel were also noted. The appendix could not be visualised separately.

DIFFERENTIAL DIAGNOSIS

Based on the history, examination findings and results of investigations, differential diagnoses of acute appendicitis, ileocaecal tuberculosis, salpingitis and tubo-ovarian abscess were considered.

TREATMENT

The patient was taken to theatre for a diagnostic laparoscopy +/- appendicectomy. At operation, a long retrocecal

Figure 1 Chest radiograph demonstrating bilateral hilar lymphadenopathy suggestive of pulmonary sarcoidosis.

Figure 2 H&E micrograph at ×10 power of granulomatous inflammation of appendix caused by sarcoidosis.

appendix was found, which appeared acutely inflamed, and was removed, along with free fluid in the pelvis. The gallbladder was distended and multiple granulomatous-looking lesions were seen in the liver. An omental mass was also found, which was subsequently biopsied on the suspicion of sarcoid granuloma.

OUTCOME AND FOLLOW-UP

Postoperatively, a repeat ultrasound was performed which showed inhomogeneity of the liver and spleen suggesting early involvement by sarcoidosis. The patient's symptoms resolved postoperatively and she was discharged home.

The appendiceal specimen confirmed granulomatous inflammation, the majority non-caseating and thus consistent with sarcoid (figure 2). A Ziehl-Neelsen stain for *Mycobacterium tuberculosis* was negative. Omental biopsy demonstrated chronic inflammatory tissue only.

DISCUSSION

Systemic sarcoidosis¹ can typically affect any organ causing granulomas to form, but predominantly affects the lungs. Common signs and symptoms include non-productive cough, fever, weight loss, chest pain, ankle swelling, lymphadenopathy, erythema nodosum and eye pain/blurred vision. Typically, sarcoidosis can be diagnosed via a combination of tissue biopsy (ie, of palpable lymph nodes) and serum ACE levels. Typical treatments include corticosteroids, immunosuppressants such as azathioprine or methotrexate, and tumour necrosis factor- α inhibitors such as infliximab.

Symptomatic appendiceal sarcoidosis, as described in the above case, is extremely rare: there are seven cases to date reported in the literature.^{2–8} Of these, three patients had a normal-looking appendix without evidence of

inflammation.^{3 4 7} The other four had inflamed appendices with appendicitis confirmed on histology, of which three perforated.^{2 5 6 8}

There are a number of differentials for granulomatous inflammation of the appendix, notably Crohn's disease, tuberculosis, histoplasmosis and *Yersinia pestis* infection. These may be indistinguishable on routine histology and further investigation may be required.⁹ Tuberculosis can be ruled out with Ziehl-Neelsen staining and histoplasmosis/*Yersinia pestis* with culture.

This is only the fourth case in the literature of appendiceal sarcoidosis causing right iliac fossa pain without evidence of appendicitis on histology. This is unusual as granulomas are often present in multiple organs in patients with systemic sarcoidosis, but usually remain asymptomatic. One review reported their presence in 75% of liver and 60% of splenic biopsies,¹⁰ and consequently, the exact cause of pain in these patients remains to be determined.

Learning points

- ▶ The clinical presentation of appendicular sarcoidosis may range from pain to perforation/abscess formation.
- ▶ There is a high rate of perforation in this group of individuals.
- ▶ Surgical intervention should not be delayed in patients with sarcoidosis presenting with right iliac fossa pain.

Competing interests None.

Patient consent Obtained.

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