

## ISOLATED ILEAL SARCOIDOSIS PRESENTING AS APPENDICITIS

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### ISOLATED ILEAL SARCOIDOSIS PRESENTING AS APPENDICITIS (ABSTRACT):

*Background:* Sarcoidosis is a granulomatous disorder of unknown etiology-with potential to involve multiple organs. The most commonly involved organs are lungs, lymph nodes and spleen. The true incidence of intestinal involvement is not known as it is very rare. *Case Report:* We report a case of a 54-year-old male with suspected sarcoidosis in remission for 10 years who presented with clinical features of acute appendicitis. On table appendix was found to be normal but terminal ileum up to ileo-caecal junction was inflamed with patchy necrosis. A limited right hemi-colectomy was done and histology confirmed sarcoidosis. Postoperative recovery was uneventful and patient remained asymptomatic until one year of follow up. *Conclusion:* Isolated terminal ileitis presenting like appendicitis in systemic sarcoidosis, especially in remission was unknown before. This case and the literature review highlights this phenomena and guide management.

KEY WORDS: SARCOIDOSIS, ILEAL SARCOIDOSIS, INTESTINAL SARCOIDOSIS, GRANULOMA, ANGIOTENSIN CONVERTING ENZYME.

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### INTRODUCTION

Sarcoidosis is a systemic disease characterized by involvement of multiple organs, specially lungs, eyes skin and joints. It is diagnosed by clinical features, radiological features of lung involvement and biopsy pictures from involved lymph nodes and lesions of noncaseating granuloma. In addition, autoantibody screen is essential to rule out other differential diagnosis. Gastro-Intestinal (GI) involvement is rare with stomach being the most commonly affected organ. In literature review, there are two-reported case of duodenal obstruction [1] and six reported cases of small intestinal involvement as the major presenting feature of systemic sarcoidosis in adults [2].

Though there are reports of appendiceal involvement in sarcoidosis, we report a rare picture where clinical diagnosis of appendicitis turned out to be an isolated ileal sarcoidosis. We also include a review the literature.

### CASE REPORT

A 54-year-old white male presented with sudden onset right iliac fossa (RIF) pain, anorexia and nausea. There were no similar history of GI symptoms in the past.

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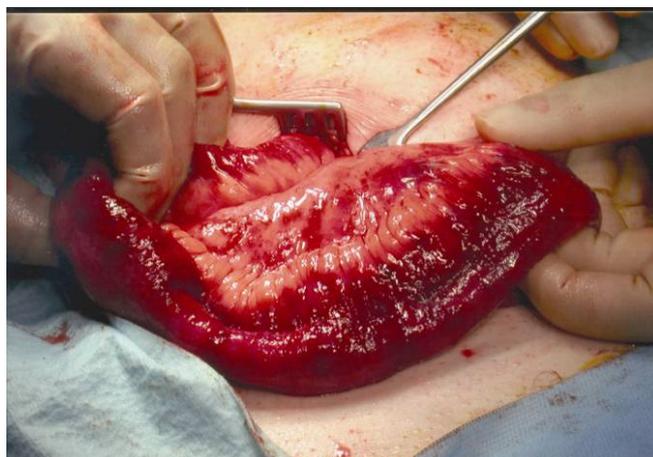
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However; he was treated almost 15 years ago for suspected sarcoidosis. His presentation that time was with low-grade fever and night sweats, respiratory symptom with cough and shortness of breath, eye symptoms with dryness and irritation, joint symptoms especially around the knees and cutaneous features like erythema nodosum. He underwent Chest X-ray which had shown emphysematous change. CT had shown bilateral pulmonary infiltrates with some precarinal lymphadenopathy. His respiratory function test showed restrictive lung pattern with reduced transfer rate. Bronchoscopy and tran-bronchial biopsies however failed to show any significant pathology and he declined any further attempt at lung biopsy. He had a normal fundoscopy. His CRP and ESR were raised (values 190 and 100 respectively) and LFT was abnormal with raised AST (151 IU/L and ALP 109 IU/L). He had a raised Serum Angiotensin converting enzyme (SACE) as well. Lyme serology done was normal. He had some occasional dyspepsia for which an endoscopy with duodenal biopsy was done which was normal. His liver biopsy showed mild nonspecific inflammatory changes. His ECG was normal. Autoantibody tests including anti nuclei, anti mitochondria, anti smooth muscle, thyroid microsomal antibody, thyroglobulin, gastric parietal cell antibody, skeletal muscle and reticulin antibody and anti-dsDNA, ANCA and RF antibody were all negative.

In view of his symptoms and raised SACE a provisional diagnosis of systemic sarcoidosis was made. He used to take oral steroid initially but he was almost symptom free for the last 10 years and without medication. His SACE and calcium, CRP and ESR repeated few years ago were normal.

On examination, there was slight pyrexia (37.7°C), pulse rate 100, localized abdominal tenderness in RIF with mild guarding and rebound tenderness. Blood examination showed leucocytosis and raised C reactive protein. A clinical diagnosis of acute appendicitis was made he was and posted for emergency appendicectomy.

At operation appendix was normal but terminal 20 cm of ileum was inflamed with patchy areas of necrosis/infracton (Fig. 1). In addition, there were few serosal and subserosal and sub mucosal firm nodules in the involved area and slight overall thickening of the segment. However, there was no sign of fat creeping or intestinal stricture suggestive of Crohn's disease. There was also mesenteric lymphadenopathy around the area. A limited right hemicolectomy was done in view of the doubtful viability of the involved ileum.



**Fig. 1** Intraoperative view - showing inflamed terminal ileum with patchy gangrene

Postoperative patient made an uneventful recovery. Histology showed minimal inflammation of appendix. The lymph node was normal. The bowel showed ischemia and focal infarction with small vessel glaucomatous vasculities and non-caseating granuloma strongly suggestive of sarcoidosis.

## DISCUSSION

Sarcoidosis is an multisystem inflammatory disease of unknown etiology characterized by the presence of non-caseating granulomas. Young to middle aged adults are commonly affected. However, there is also a peak in incidence after 40 years of age.

It requires the presence of involvement in two or more organs for a specific diagnosis. The patients usually present with bilateral hilar lymphadenopathy, pulmonary infiltrates, and ocular and skin lesion.

Diagnosis is based on clinical and radiologic finding's plus histologic evidence of non-caseating epithelioid granuloma, with exclusion of other granulomatous diseases.

Recent theories point to an environmental agent, either infectious or non-infectious that triggers inflammation in genetically susceptible host. Mycobacterial and fungal infections, malignancy and environmental agents such as beryllium are commonly implicated. There is proof of association with *Propionibacter acnes* and with mKatG protein found in mycobacterium.

The clinical outcome of sarcoidosis varies, with remission occurring in over half the patients within a few years of diagnosis; however, the remaining patients develop a chronic disease that lasts for decades.

Laboratory investigations include SACE levels. Angiotensin-converting enzyme is produced by the epithelioid cells of the granuloma and reflects total body granuloma load. However, it can have a false-positive rate of 10% and a false-negative rate of 40%. Our patient had an elevated SACE, which responded to steroid along with a negative autoantibody screen supporting the diagnosis of sarcoidosis.

X ray and CT findings such as with bilateral hilar lymph node involvement with or without pulmonary fibrosis and infiltrate are also suggestive. CT guided biopsy and bronchoscopy with biopsy helps in confirmation of the diagnosis. In our case CT finding were suggestive though biopsy attempt was unsuccessful.

Though we have mainly limited our discussion to sarcoidosis of ileum but we also reviewed different manifestations of small intestinal sarcoidosis.

All published case reports were identified by comprehensive electronic search of Medline database (1966-2009). A search for medical subject heading ileal and small intestinal and keyword search for sarcoidosis and granulomatous disease was performed. This strategy was supplemented by a manual search of the bibliographies from all retrieved publications.

Intestine is rarely found involved in sarcoidosis. In early reports [3] diagnosis was based on presence of non-caseating granuloma in the intestine and mesenteric lymph nodes without evidence of pulmonary and systemic disease. Maycock found no case report of intestinal sarcoidosis in his literature review in 1963 [4]. However there are reports of GI sarcoidosis involving from oesophagus to large bowel.

Stomach is the most frequently involved organ in intestine. Gastric sarcoidosis, particularly involving the antrum, affects approximately 10% of patients with systemic disease. GI sarcoidosis commonly occur subclinically, with clinical manifestations present in only 0.1 to 0.9% of patients with the disease [5].

Sarcoidosis of small intestine can present with diverse clinical signs and symptoms. Protein losing enteropathy [6] or hemorrhage can be the first symptom. Bleeding from sarcoidosis of the esophagus, stomach, and colon has also been reported. It can present as a granulomatous enterocolitis resembling Crohn's disease [7]. There are case reports of disseminated GI sarcoidosis and also rectal and large bowel involvement and mesenteric lymph node involvement, simultaneous involvement of duodenum and ileum [8].

Isolated sarcoidosis of the ileum can cause of acute intestinal obstruction [9]. Carcinoma of the terminal ileum along with evidence of sarcoidosis of ileum is reported in ulcerative colitis [10]. There are few reports of Crohn's disease of small intestine and colon turning out to be sarcoidosis but isolated terminal ileitis in a case of sarcoidosis mimicking appendicitis was unknown before.

Some possible clinical association between celiac disease and sarcoidosis and folate deficiency has been found. One paper has demonstrated a moderately increased prevalence of celiac disease in Irish patients with sarcoidosis, which they feel justifies future screening of sarcoid population [11]. Another study has demonstrated a high frequency of gastric autoimmunity and gluten-associated immune reactivity in patients with sarcoidosis, occurring in almost 40% of the cases, the former being the most frequent gastrointestinal immune manifestation. Overall, despite a high frequency of humoral autoimmunity the frequencies of clinical disease, pernicious anaemia and celiac disease were not increased as compared with the control population. However since our patient did not had any GI symptom for the last 10 years so this possibility was not separately explored.

There are cases reported, albeit very few, where initial manifestations suggested acute appendicitis which later turned out to be appendicular involvement of sarcoidosis. On further investigations systemic sarcoidosis was identified in them [12]. However, there is no report of presentation mimicking acute appendicitis but turning out to be ileal sarcoidosis.

## CONCLUSION

Isolated terminal ileal involvement presenting like acute appendicitis was unknown before though there are reports if small intestinal obstruction. Our report highlights possibility of flare up of sarcoidosis after a long time and that it could involve an entirely new system uninvolved before. Although for an isolated terminal ileitis is usually treated conservatively, but in our case, patchy necrosis tilted the decision in favor of resection. Amongst varied differential diagnosis of appendicitis, ileal sarcoidosis should also be included as one in a case where there is a previous history of sarcoidosis.

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