

# Gastric Sarcoidosis: A Difficult to Diagnose Rare Disease

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**Abstract** Gastrointestinal sarcoidosis is a rare clinical entity. Diagnosis of isolated gastric sarcoidosis is difficult as it is usually asymptomatic; when symptomatic it presents with non-specific symptoms such as abdominal pain, nausea and vomiting. We here present a case of a 32-year-old black lady who presented with non-specific abdominal complaints; a diagnosis gastric sarcoidosis was established following endoscopic biopsy. Here symptoms resolved promptly with steroidal therapy as with most cases. Gastric sarcoidosis should be suspected in sarcoid patients who present with nonspecific abdominal complaints. This case serves as an important clinical reminder of the atypical manifestations of sarcoidosis.

**Keywords:** *gastric sarcoidosis, sarcoidosis, esophagogastroduodenoscopy, corticosteroidal therapy*

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## 1. Introduction

Sarcoidosis is a multi-organ granulomatous disorder of unknown etiology. 90% of the patients with sarcoidosis present with intrathoracic lymphadenopathy, pulmonary involvement, skin and ocular symptoms and signs or any combination of the above. Clinical presentation with gastrointestinal (GI) symptoms is seen in 0.1-0.9% of patients with systemic sarcoidosis [1]. The stomach (antrum) is the most affected organ in GI sarcoidosis [2]. Though most cases of gastric sarcoidosis remain asymptomatic, patients can present with nonspecific signs and symptoms such as epigastric pain, anorexia and vomiting [3]. Isolated gastrointestinal sarcoidosis may pose a diagnostic challenge. Diagnosis is established with upper GI endoscopy with mucosal biopsies. Histopathology regularly demonstrates noncaseating granulomas, albeit other causes of granulomatous gastritis must be ruled out. We present a case of a 32-year-old black female who presented with epigastric pain and significant weight loss, who was misdiagnosed with bulimia and GERD, was subsequently diagnosed with gastric sarcoidosis, after all other causes were excluded. A detailed discussion regarding clinical features, workup and histological findings in gastric sarcoidosis follows. This case serves as an important clinical reminder of the atypical manifestations of sarcoidosis.

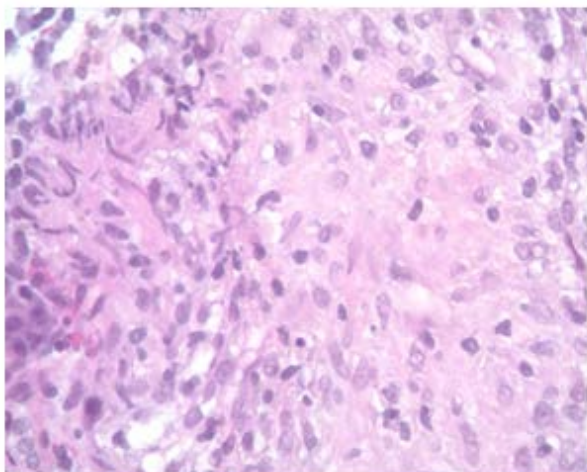
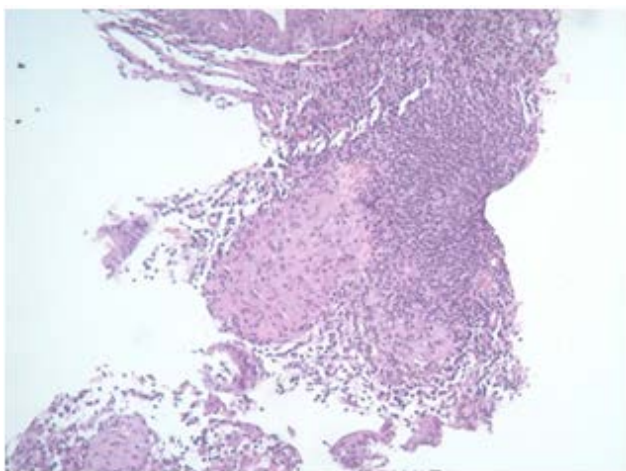
## 2. Case Presentation

A 32-year-old- black female presented with epigastric pain, difficulty swallowing, bloating sensation after eating, persistent vomiting and weight loss for the past year. She was clinically diagnosed to have bulimia nervosa and gastro-esophageal reflux disease; the latter was treated with esomeprazole (40 mg, orally, once daily) without much benefit. Physical examination was significant for cachexia with a body mass index of 14.9 kilogram per meters square, and poor dentition with loss of many teeth. Cardiopulmonary and abdominal examination was normal. Laboratory investigations were significant for microcytic hypochromic anemia with a hemoglobin level of 9.6 g/dl, total white blood cell count of 3900/microliter, lymphopenia at 16% and serum albumin of 3.3g/dL; erythrocyte sedimentation rate and C-reactive protein were within normal limit. Chest radiography was normal. As symptoms of weight loss persisted despite medical management for gastro-esophageal reflux disease, an esophago-gastric-duodenoscopy was performed which revealed nodular gastritis (Figure 1) and a hiatal hernia. Histological examination of a biopsy specimen from the gastric antrum revealed moderate to severe chronic active gastritis with foveolar hyperplasia and a lymphoid aggregate. Gastric body biopsy revealed moderate chronic active gastritis with non-caseating granulomas and lymphoid aggregates (Figure 2). No *Helicobacter pylori*, acid fast bacilli or fungi were seen on giemsa stain, acid fast stain (Figure 3) and fungal stains respectively. Acid-fast stain and culture of the gastric aspirate were negative. Interferon gamma assay was negative and angiotensin converting enzyme levels obtained was within normal limit. Thus a diagnosis of gastric sarcoidosis was established. Medical management with prednisone (50 mg,

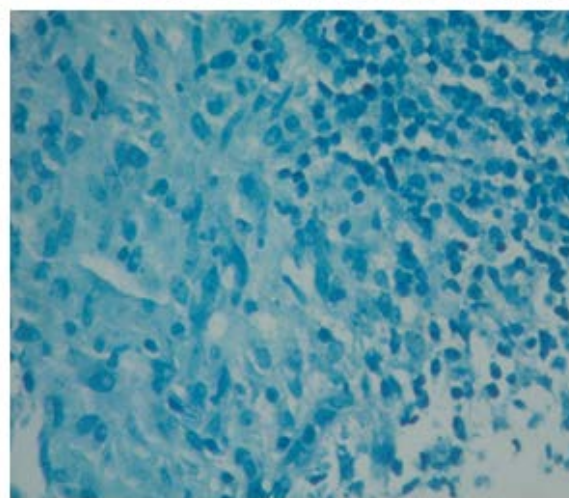
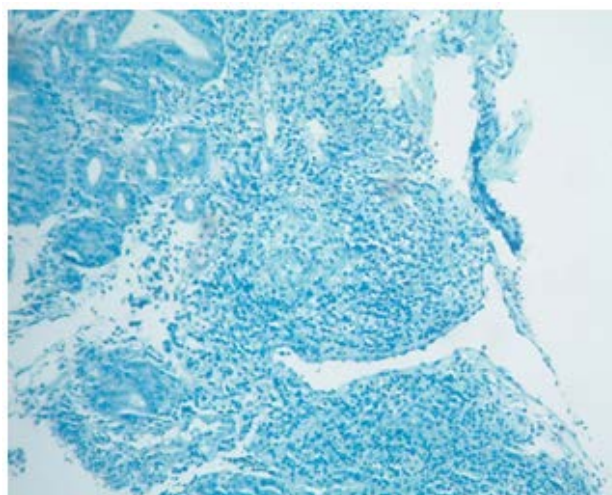
orally, once daily) was started with omeprazole (20 mg, orally, once daily). The patient, initially noncompliant to medical management subsequently became compliant; cosyntropin (80 units, subcutaneously, once every three days) added to the treatment led to improvement in symptoms. Pulmonary function test performed subsequently revealed FEV1/FVC (forced expiratory volume 1/ forced vital capacity) of 115 and decreased diffusion capacity of lungs for carbon monoxide (DLCO) consistent with restrictive lung disease of sarcoidosis.



**Figure 1.** Gastric endoscopy revealing nodular sarcoid lesions in stomach



**Figure 2.** Hematoxylin and eosin staining of gastric biopsy showing non-caseating granuloma and mucosal lymphocyte infiltration



**Figure 3.** Acid fast stain of gastric biopsy that revealed absence of acid fast bacillus

### 3. Discussion

Sarcoidosis is a systemic disease of unknown etiology characterized by the formation of noncaseating granulomas in the organs affected. Sarcoidosis commonly affects intrathoracic lymph nodes, lungs, skin and eyes [1]. Gastrointestinal sarcoidosis is rare and may happen alone or with pulmonary and other organ involvement. the stomach, particularly the antrum is the most commonly affected organ in gastrointestinal sarcoidosis [2]. Clinically recognizable gastric sarcoidosis is noted in < 1% of sarcoidosis patients [4]. Israel et al report a 0.6 % incidence of symptomatic gastrointestinal sarcoidosis in a study of 160 sarcoid patients [5].

In an analysis of 44 biopsy-proven gastric sarcoid cases, epigastric pain and emesis were reported to be the most common symptoms, as noted in 70% and 33% of cases respectively [3]. Other symptoms include early satiety, vomiting, hematemesis, melena and weight loss [6]. Very rarely gastric sarcoidosis may present as pernicious anemia and irritable bowel syndrome [7].

Laboratory investigations are not helpful in the diagnosis of gastric sarcoidosis as they may reveal nonspecific abnormalities such as peripheral lymphopenia with CD4 T-lymphocyte depletion, elevated levels of serum angiotensin converting enzyme (ACE), lysozyme,

beta-macroglobulin, hypercalcemia and hypercalciuria [8]. Gastric aspirate may show elevated ACE levels [9].

Esophagogastroduodenoscopy is required to obtain a biopsy that may reveal noncaseating granulomas. It is important to note that, 50% of the gastric sarcoid cases have no visible lesion on endoscopy [3] and non-caseating granulomas may be absent in biopsies. Palmer et al followed 6 patients with biopsy confirmed gastric sarcoidosis for a period of 18 months, during which none of the patients became symptomatic though there were no change in biopsy patterns [10]. One or more of the following endoscopic findings may be noted in gastric sarcoidosis : i) Single or multiple ulceration that may mimic peptic ulcer disease [11], ii) cone shaped antral deformities [12], iii) polyposis [13], iv) diffuse nodular lesions [14], v) linitis plastica like lesion (irreversible) [13,15,16]. Other endoscopic abnormalities noted may include gastric wall thickening [17,18,19,20], cobble stoning [11,12,13,14,21] and pyloric stenosis [22]. Gastric biopsy are subjected to various stains to rule out other granulomatous disorders such as crohn's disease, malignancy, foreign body, tuberculosis, parasites, bacteria, fungi, syphilis and helicobacter pylori [23]. Endoscopic ultrasonography and computed tomography of the abdomen may show other abdominal sarcoid manifestations such as regional and retroperitoneal lymphadenopathy and hepatosplenomegaly [9,24,25,26].

There is no established standardized treatment for the management of gastrointestinal sarcoidosis. Based on recommendations for the management of gastrointestinal sarcoidosis corticosteroids are the initial drug of choice. Asymptomatic cases do not require treatment and most symptomatic cases of gastrointestinal sarcoidosis respond to oral prednisone at 20-40 mg per day. Steroid refractory cases are managed with antimetabolites such as methotrexate, azathioprine and leflunomide; tumor necrosis factor alpha antagonists are used as third line agents [23,27]. Gastric resection is considered in cases that are refractory to medical management, obstruction, severe malabsorption or hemorrhage [28].

## 4. Conclusion

Gastrointestinal sarcoidosis is a very rare manifestation of sarcoidosis that may occur as a part of multiorgan involvement or in isolation. Stomach (antrum) is the most commonly affected region in gastrointestinal sarcoidosis. Due to nonspecific signs and symptoms diagnosis of gastric sarcoid is often difficult. In patients with established sarcoid patients who develop non-specific gastrointestinal symptoms, suspicion for gastrointestinal sarcoidosis should be high. Endoscopic biopsy is the key to establish the diagnosis of gastric sarcoidosis as it not only reveals non-caseating sarcoid granulomas, but also rules out other granulomatous diseases. Though no established guidelines exist for the management of gastrointestinal sarcoidosis, majority of cases promptly respond to oral steroids.

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