CASE REPORT

Isolated gastric sarcoidosis: a rare entity

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SUMMARY

We present a case of isolated granulomatous gastritis in a 21-year-old woman. Initial symptoms included nausea, vomiting and inability to tolerate oral intake. An upper oesophagogastrroduodenoscopy revealed nodular and thickened mucosa with histological findings of granulomatous gastritis. Infectious, inflammatory and malignant causes were excluded prior to making a diagnosis of gastric sarcoidosis.

BACKGROUND

Granulomatous gastritis is a rare entity with low incidence.¹ Granulomatous gastritis can be seen in patients with a variety of conditions including Crohn's disease, sarcoidosis and Whipple's disease. It is also seen in certain infections such as *Helicobacter pylori* and tuberculosis and malignancies including lymphoma and adenocarcinoma.^{1 2} Gastric sarcoidosis is uncommon and is found in only 2.5% of autopsy cases of individuals with a diagnosis of systemic sarcoid.³ Intestinal sarcoidosis in general is also rare, affecting only 3.4% of patients with systemic sarcoid.³ Sarcoidosis without pulmonary involvement is also uncommon, affecting less than 10% of patients.⁴ Current literature suggests isolated gastric sarcoidosis as a potential clinical entity; however, a review of case reports on the topic only describes gastric sarcoidosis in combination with extraintestinal disease.^{5 6} Here, we present a case of isolated granulomatous gastritis secondary to sarcoidosis without extraintestinal manifestations.

CASE PRESENTATION

A 21-year-old Caucasian woman with no past medical history presented to the emergency department with nausea, vomiting and inability to tolerate oral intake. She noted symptoms over the past month, initially beginning with midline chest discomfort, described as burning with radiation to the back. Soon after, she began experiencing postprandial nausea, vomiting and abdominal discomfort. Prior to her hospital presentation, she had been assessed at two other institutions and had been prescribed proton pump inhibitors and antiemetics for symptom control without improvement.

She had used aspirin containing over-the-counter medication for migraine and up to 9 units of alcohol occasionally. However, she had stopped consuming alcoholic beverages since the onset of her symptoms. In the month prior to presentation at our institution, she developed three painful nodules on

her thigh, which had resolved by the time of presentation.

Five years prior to this illness, a biopsy of an enlarged right inguinal lymph node showed florid reactive hyperplasia and necrotising granulomatous lymphadenitis with stellate abscesses and with morphological appearance consistent with cat scratch disease.

Her initial observations revealed a temperature of 36.1°C, heart rate 101 bpm, respiratory rate 20/min, blood pressure 116/75 mm Hg and oxygen saturation 100% on room air. Physical examination of her cardiovascular and respiratory systems was unremarkable. Abdominal examination demonstrated tenderness on palpation of the epigastrium without palpable masses or hepatosplenomegaly. She had no rash nor signs of synovitis.

INVESTIGATIONS

Complete blood count revealed a mild microcytic anaemia. Liver function tests and lipase were normal. Calcium was 9.4 mg/dL (normal range 8.8–10.7). Erythrocyte sedimentation rate was mildly elevated at 23 mm/hour. C-reactive protein was elevated at 88 mg/L. Serum *H. pylori* testing returned positive. Given lack of prior treatment for *H. pylori*, stool antigen testing and breath testing were not additionally pursued. T-spot testing for tuberculosis was negative. Infectious studies for histoplasmosis (*Histoplasma* serum antigen negative), toxoplasmosis (*Toxoplasma* IgM and IgG antibody negative), coccidiosis (*Coccidioides* antibody negative), syphilis (treponemal IgG antibody negative), *Cryptococcus* (cryptococcal antigen negative), *Aspergillus* (galactomannan antigen 0.13 (normal 0–0.49)), *Bartonella* (*B. henselae* IgM antibody titre <1:20, *B. quintana* IgM antibody titre <1:20) and schistosomiasis (*Schistosoma* Ab <1.00 (normal 0–0.99)) were negative. Testing for chronic granulomatous disease with neutrophil oxidative burst assay was negative. Serum ACE level was elevated at 68 U/L (normal range 8–53).

Chest imaging including plain radiographs and CT was normal without parenchymal lesions or hilar/mediastinal lymphadenopathy. Pulmonary function tests, including spirometry, lung volumes and diffusion capacity for carbon monoxide were normal.

Additional workup during her hospitalisation included an abdominal ultrasound that was notable for mesenteric lymphadenopathy, measuring up to 1.7 cm, and a MRI of the abdomen that showed diffuse gastric wall thickening and submucosal enhancement in the stomach with perigastric stranding, upper abdominal lymphadenopathy

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Figure 1 Top left: endoscopic ultrasound showing thickened gastric wall. Top right: abdominal MRI showing increased T2 signal within the submucosa of the gastric body and antrum. Bottom left: coronal T2-weighted image demonstrating abnormal T2 signal within submucosa of gastric body/antrum. Bottom right: axial T2 sequence demonstrating abnormal perigastric and omental lymph nodes (arrows).

and omental nodules (figure 1). Duodenum and small bowel appeared normal.

An upper oesophagoduodenoscopy (OGD) was performed revealing diffusely nodular and thickened mucosa with a rigid wall along the entire lesser curvature of the stomach (figure 2). Colonoscopy was unremarkable. Additional intestinal evaluation with video capsule endoscopy revealed normal small bowel.

Gastric biopsies revealed severe chronic active granulomatous gastritis (figure 3). Additional studies from her gastric biopsies, including gram stain, fungal stain and acid-fast stain, were negative. Biopsies from the colon and terminal ileum were normal. The patient also underwent fluoroscopic-guided biopsy of a perigastric lymph node with findings consistent with a reactive process. Stains for pathogens and cultures of the biopsy were negative. Biopsies did not reveal signs of malignancy.

Figure 2 Upper endoscopy revealing diffusely nodular and thickened mucosa with a rigid wall.

DIFFERENTIAL DIAGNOSIS

A broad differential was initially considered including infectious aetiologies, Crohn's disease, sarcoidosis, chronic granulomatous disease and malignancy. Infectious differential included *H. pylori*, tuberculosis, histoplasmosis, toxoplasmosis, coccidiosis, syphilis, *Cryptococcus* and schistosomiasis.

TREATMENT

Following initial hospitalisation, the patient was started on budesonide and esomeprazole for possible Crohn's disease. However, given the ambiguity regarding diagnosis, the budesonide was stopped after 1 month with persistent symptoms, while full infectious workup was pursued. Serum *H. pylori* testing returned positive, and the patient completed a course of triple therapy with amoxicillin, clarithromycin and omeprazole. Although a positive serology does not always indicate active infection, the patient was treated for *H. pylori* infection given lack of prior treatment and the possibility of contribution to her symptoms.⁷ Breath testing following antibiotics for *H. pylori* was negative; however, symptoms persisted. She eventually was treated with prednisone (40 mg daily for 1 week with a taper down by 10 mg weekly for a total of 4 weeks) with minimal clinical improvement. Given concern that the initial duration of high-dose steroids was not sufficient, she received an additional course of prednisone (40 mg daily for 2 weeks with a taper of 10 mg each week). Following the second course of steroids, the patient noted reduced frequency and improvement of symptoms and OGD showed endoscopic improvement (figure 4).

OUTCOME AND FOLLOW-UP

Corticosteroids were slowly tapered over 5 weeks and a marked improvement of symptoms was reported prior to completing the second course of prednisone. Acid suppression with omeprazole 20 mg daily for reflux was also used and currently the patient is asymptomatic. She remains on a proton pump inhibitor and is being monitored by rheumatology and gastroenterology.

Figure 3 Gastric mucosa with granulomas.

DISCUSSION

Granulomatous gastritis is a rare entity with the differential including infectious, inflammatory and malignant causes. With regard to the inflammatory aetiologies, Crohn's disease is one of the most common causes of granulomatous gastritis.² Crohn's disease was considered here, but ultimately felt to be less likely given the lack of other small bowel or colonic involvement as well as lack of characteristic pathology. Although rare, sarcoidosis involvement of the gastrointestinal tract does occur and most frequently involves the stomach.^{8,9}

Sarcoidosis is typically a multiorgan disease with histological presence of granulomas, making its recognition by providers of various specialties important. Sarcoidosis is usually diagnosed in adulthood and a nationwide register-based study in Sweden found a peak incidence in women between 50 and 60 years old.¹⁰ Characteristically, sarcoidosis is found within the thoracic cavity with lung involvement in over 90% of patients.⁸ Isolated extrapulmonary sarcoidosis is rare with one cohort study describing this disease pattern in only 3% of patients.¹¹ Gastric sarcoidosis has previously been reported in the literature, though usually with extraintestinal manifestations.^{5,9} While many studies report gastrointestinal involvement in patients with a known history of sarcoidosis, gastric sarcoidosis has also been reported as the initial site of organ involvement but later other foci of disease activity have been identified.⁶ A review of 44 cases of patients with gastric sarcoidosis showed epigastric pain and emesis to be the two most common associated symptoms, while early satiety manifested in patients with more infiltrative disease.⁶

We present a case of isolated gastric sarcoidosis without extraintestinal manifestations. It is unclear if the previous lymphadenopathy occurring 5 years prior to presentation was related to our patient's current diagnosis though history and histological features at that time favoured an infectious aetiology. Although sarcoidosis typically presents as a systemic disease with multiple sites of involvement, our patient has not yet had obvious extraintestinal organ involvement of sarcoidosis apart from abdominal lymphadenopathy and possible erythema nodosum.¹² Her history of developing three painful, erythematous lower limb lesions was suggestive of

erythema nodosum but this resolved spontaneously prior to her presentation so could not be formally confirmed. Given her clinical presentation, granulomas on gastric biopsy, negative evaluation for other diseases, prior and current lymphadenopathy, and elevated serum ACE level, a diagnosis of gastric sarcoidosis was favoured.

As sarcoidosis is typically a multiorgan disease, evaluation for pulmonary involvement, the most common organ involved with sarcoid, with chest imaging and pulmonary function testing was performed and negative. Salivary gland biopsy as an additional diagnostic tool was not pursued here given historical low diagnostic yield as well as higher positivity among patients with hilar lymphadenopathy.^{13,14} Positron emission tomography imaging can assist with diagnosing unexplained, persistent symptoms when there is a high suspicion for sarcoidosis; however, such imaging was not obtained given explanation of the patient's symptoms by endoscopic and histological findings.¹⁵ It is possible our patient may develop additional organ involvement and she will require close monitoring and evaluation should other symptoms arise.

Our patient initially presented with nausea, vomiting, and inability to tolerate oral intake with diagnostics most notable for nodular and thickened mucosa on OGD and severe chronic active granulomatous gastritis on histology. Gastric sarcoidosis, given its low incidence, and the lack of involvement in other organs, became a diagnosis of exclusion in our case. The broad differential of granulomatous gastritis required an interdisciplinary approach to exclude other possible and more common aetiologies. Our patient remains well and is under continued outpatient clinical review.

Learning points

- ▶ Granulomatous gastritis is a rare entity with low prevalence and many potential infectious, inflammatory and malignant aetiologies.
- ▶ Although gastrointestinal sarcoidosis is rare, the stomach is the most commonly involved site.
- ▶ Gastric sarcoidosis generally presents with extraintestinal manifestations; however, isolated gastric sarcoidosis does exist and may be the initial presentation of sarcoidosis.

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Figure 4 Improved endoscopic appearance of the gastric antrum after treatment with steroids.

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