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CASE REPORTS

Necrotizing Sarcoid Granulomatosis: A Case Report of Gastric Involvement

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Necrotizing sarcoid granulomatosis (NSG) is an immune system disorder characterized by necrotizing granulomas, as opposed to noncaseating granulomas in classical sarcoidosis. Over the past 3 decades there have been over 120 reported cases of NSG with pulmonary and extrapulmonary involvement.[1] We present a patient found to have histological evidence of necrotizing granuloma in her gastric antrum, and we believe this is the first Case Report of NSG involving the stomach.

Case Report

A 21-year-old African-American female first presented to an outside hospital with fever, epigastric pain, shortness of breath, and headache. Two days later she complained of nonbloody, nonbilious vomiting, and was found to have leukocytosis (17,770 cells/m^3), elevated lipase (224 U/L), elevated C-reactive protein (14.7 mg/L), and an inflamed pancreas on computed tomography (CT). She was treated conservatively for pancreatitis and started on ampicillin/sulbactam. After 2 contrast CT scans on consecutive days, she developed acute renal failure (creatinine 2.0 mg/dL compared to baseline of 1.0 mg/dL), and was transferred to our hospital for further evaluation and management.

Upon transfer, the patient's temperature was 37.0°C, pulse was 102 beats/minute, blood pressure was 141/84 mm Hg, and oxygen saturation was 94%. On examination, she was tender to palpation in her epigastrium and right upper quadrant, but the remainder of the physical exam was unremarkable. She was started on moxifloxacin and managed with intravenous (IV) fluid hydration and pain control. Within 3 days, CT showed resolving pancreatitis, magnetic resonance cholangiopancreatography (MRCP) was negative, and her creatinine began normalizing (1.3 mg/dL). Nonetheless, she continued to complain of abdominal pain, shortness of breath, and intermittent low-grade fevers. She then also developed bilateral panuveitis requiring high-dose steroid eye drops.

Chest x-ray showed subtle bilateral nodular and bronchiolitic infiltrates with no evidence of enlarged hilar nodes, and subsequent bronchoscopy showed no abnormalities. Additional workup included negative blood and urine cultures, purified protein derivative (PPD), and *Clostridium difficile* assay; as well as negative human immunodeficiency virus (HIV), cryptococcus, *Helicobacter pylori*, *Borellia burgdorferi*, syphilis (fluorescent treponemal antibody), aspergillus, histoplasma, and rheumatological serologies. Her white blood count (20,900 cells/m^3), C-reactive protein (6.6 mg/L), and erythrocyte sedimentation rate (100 mm/hour) remained elevated.

The patient continued to complain of epigastric pain. Repeat abdominal CT scan showed large retroperitoneal and mesenteric lymph nodes, and esophagogastroduodenoscopy (EGD) showed gastritis with an antral nodule (Figure 1). Biopsy of the nodule revealed a necrotizing granuloma with mixed cellular infiltrate. Biopsy stains were negative for bacteria, borellia, treponemes, acid-fast bacilli, and fungi. The patient was diagnosed with necrotizing sarcoid granulomatosis and started on an oral prednisone taper. She responded to steroid treatment with prompt resolution of her uveitis, shortness of breath, abdominal pain, and fevers. She was discharged following treatment, has continued to do well, and is seen regularly at the sarcoid clinic for follow-up.

Discussion

NSG was first characterized as a distinct variation from sarcoidosis by Liebow[2] in 1973, and was noted to have 3 characteristic differences: (1) histological evidence of sarcoid-appearing granuloma and necrosis, (2) pulmonary nodules without hilar lymphadenopathy on imaging, and (3) a clinically benign course. Over the past 3 decades, there have been more than 120 cases of reported NSG involving the lungs, gastrointestinal tract, kidney, skin, and central nervous system.[1] Since Liebow's[2] description, newly reported NSG cases have generally been found to be consistent with the aforementioned criteria, although hilar lymphadenopathy may be particularly more common than previously thought. One review noted a range of 8% to 79% prevalence

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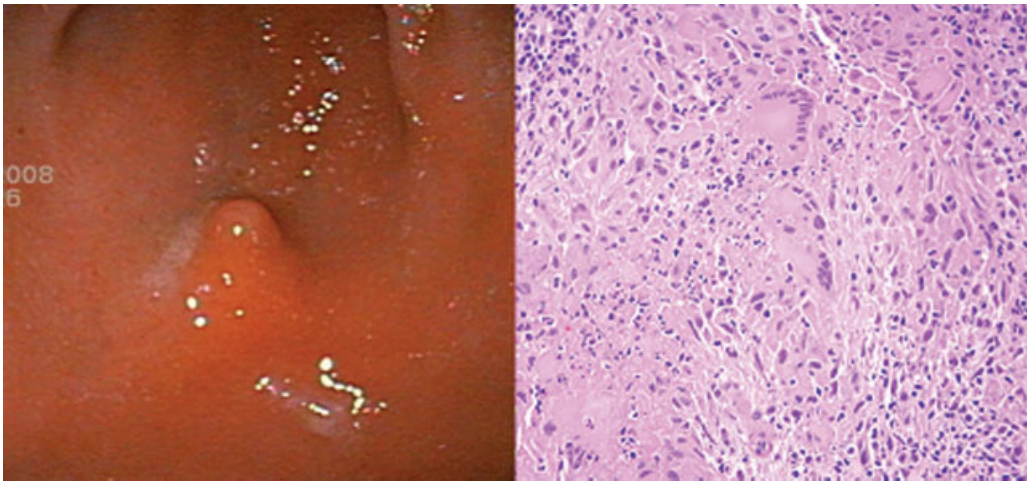

FIGURE 1. (Left) Antral nodule on esophagogastroduodenoscopy (EGD). (Right) Hematoxylin and eosin (H&E) stain (400×) of antral nodule, showing necrotizing granuloma with mixed cellular infiltrate. **Abbreviation**: H&E; hematoxylin and eosin.

of hilar lymphadenopathy in reported NSG series.[3] Therefore, while hilar lymphadenopathy still currently appears to be less common in NSG than in typical sarcoidosis, its presence should not rule out the diagnosis.

Our patient's history is consistent with Liebow's[2] criteria since she had histological evidence of necrotizing granuloma and pulmonary involvement without hilar lymphadenopathy, and responded promptly to steroid treatment. To our knowledge, this is the first case of NSG reported in the stomach.

While less than 1% of sarcoid patients are reported to have gastrointestinal involvement, all of these cases have either been noncaseating granulomas (classical sarcoidosis) or were found outside the stomach.[1,4,5] Most of the data regarding sarcoid symptomatology and treatment are derived from reports on classical sarcoidosis. In classical sarcoidosis, there is gastric antral involvement in approximately 10% of patients with systemic disease.[4] These patients may present with nausea, vomiting, and weight loss, and are often effectively treated with a single dose of prednisone 30 to 40 mg followed by a maintenance dose of 10 to 15 mg daily over 6 months.[5] Less data are available regarding necrotizing sarcoid presentation and treatment, especially with regard to gastrointestinal involvement.

We hope to raise awareness regarding: (1) the variation in noncaseating versus necrotizing sarcoid-type disorders, (2) the benefit of steroid treatment once infectious etiologies are ruled out, and (3) the potential for further extrapulmonary involvement in previously unreported organ systems.

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References

1. Strickland-Marmol LB, Fessler RG, Rojiani AM. Necrotizing sarcoid granulomatosis mimicking an intracranial neoplasm: clinicopathologic features and review of the literature. *Mod Pathol.* 2000;13(8)909–913.
2. Liebow AA. Pulmonary angiitis and granulomatosis. *Am J Respir Dis.* 1973; 108:1–18.
3. Frazier AA, Rosado-de-Christenson ML, Galvin JR, Fleming MV. Pulmonary angiitis and granulomatosis: radiologic-pathologic correlation. *Radiographics.* 1998;18(3):687–710.
4. Fireman Z, Sternberg A, Yarchovsky Y, et al. Multiple antral ulcers in gastric sarcoid. *J Clin Gastroenterol.* 1997;24(2):97–99.
5. Friedman M, Ali MA, Borum ML. Gastric sarcoidosis: a case report and review of the literature. *South Med J.* 2007;100(3):301–303.

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