CLINICAL VIGNETTE

Esophageal Sarcoidosis

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Introduction

Sarcoidosis is an inflammatory disease of unknown etiology that can affect almost any organ system. While most commonly causing pulmonary manifestations, sarcoidosis occasionally affects the gastrointestinal tract. The following is a rare case of a patient with an esophageal stricture caused by sarcoidosis.

Case Summary

A 55-year-old African American woman with gastroesophageal reflux disease presented to the hospital with several months of abdominal pain and food intolerance. The patient did not have any significant dermatologic, pulmonary, or lymphatic findings on physical exam. Endoscopy demonstrated multiple esophageal and duodenal ulcerations and an esophageal stricture requiring stent placement. Biopsy of the stricture demonstrated esophagitis along with candida and the cardia mucosa showing foveolar cell hyperplasia. Following esophageal stent placement and proton pump inhibitors, the patient only had mild improvement of her food intolerance.

The patient had several prolonged admissions for worsening nausea and emesis with nutritional deficiencies over the subsequent two years. She received brief total parenteral nutrition support, multiple esophageal stent placements, and discussion of possible esophagectomy. She was also diagnosed with non-ischemic systolic heart failure of unclear etiology. Her heart failure was suspected to be related to nutritional deficiencies from her esophageal stricture. Because of refractory symptoms from her esophageal stricture, she eventually underwent an Ivor Lewis esophagectomy. Surgical Pathology of the excised distal esophagus and stomach demonstrated noncaseating granulomas consistent with sarcoidosis.

When interviewed further, the patient denied any family or prior history of sarcoidosis and denied symptoms of pulmonary disease or other extrapulmonary disease. She had evidence of subclinical pulmonary disease with pulmonary reticular opacities and calcifications on CT thorax. However, CT showed no hilar adenopathy. Rheumatology was consulted and steroid treatment was deferred. She was discharged home on J-tube feeds with subsequent normalization of her weight and oral intake and eventual J-tube removal. She remains free of any new clinical manifestations of sarcoidosis.

Discussion

Sarcoidosis is a systemic granulomatous disease with an unknown cause. The annual incidence of sarcoidosis among adults varies across region and ethnic groups. Studies report incidence of 35.5 per 100,000 for African Americans and 10.9 per 100,000 for Caucasians.[1] The most common sites of involvement are the lungs and lymph nodes. Additional organ systems that can occasionally be involved include the eyes, skin, and muscles. Rarely there can be neurologic, cardiac, or gastrointestinal manifestations. Biopsy of sarcoid affected tissues will demonstrate noncaseating granulomas.

Gastrointestinal tract involvement of sarcoidosis is rare. Clinically significant gastrointestinal tract involvement has been reported in 0.1% to 0.9% of patients with sarcoidosis, though in some populations this may be higher.[2] For example, a Japanese study found gastrointestinal involvement in 1.6% of patients with sarcoidosis.[3] When sarcoidosis involves the gastrointestinal system, the stomach is most commonly involved. However, sarcoidosis can affect all parts of the gastrointestinal tract including the esophagus, small intestine, colon, pancreas, and peritoneum. Esophageal sarcoidosis is extremely rare.

Esophageal involvement can present with dysphagia, weight loss, abdominal pain, odynophagia, dysphonia, and anemia.[4] Endoscopic evaluation may reveal ulcerations and strictures. Esophageal involvement can be divided into four categories: superficial mucosal, esophageal musculature, myenteric, or extrinsic compression. Superficial mucosal involvement can cause mucosal hyperemia, nodular lesions, plaque-like lesions, or stricture. Esophageal musculature involvement can lead dysphagia and is usually found on biopsy showing the presence of noncaseating granulomas. Sarcoidosis of the enteric plexus can also result in dysphagia and a clinical picture similar to achalasia. Finally, lymphadenopathy can cause extrinsic compression of the esophagus leading to dysphagia.[4]

Treatment

Due to rarity of esophageal sarcoidosis, no published clinical trials compared different treatments. Current treatments include proton pump inhibitors, surgical resection, corticosteroids, and dietary modification. Surgery can relieve obstruction caused by esophageal sarcoidosis. Current surgical options include laparoscopic Heller's myomectomy with partial fundoplication or esophagectomy.[5] Corticosteroid treatment in esophageal sarcoidosis has not been well studied, however oral corticosteroids

are used occasionally, especially with systemic involvement. One review reports esophageal inflammation often responds to oral corticosteroids.[6]

Conclusion

This patient presented with clinically significant gastrointestinal disease with an initial biopsy negative for sarcoidosis. It is important to rule out alternative causes of esophageal strictures and other disease processes that can cause granulomatous disease. Our patient did well after surgical resection and has tolerated a regular diet. She has not developed recurrent symptoms or further complications from her sarcoidosis. Though rare, sarcoidosis with primarily gastrointestinal clinical manifestation does exist and can cause significant morbidity. Gastrointestinal sarcoidosis is a rare manifestation of a common disease with limited evidence to guide management.

Figures

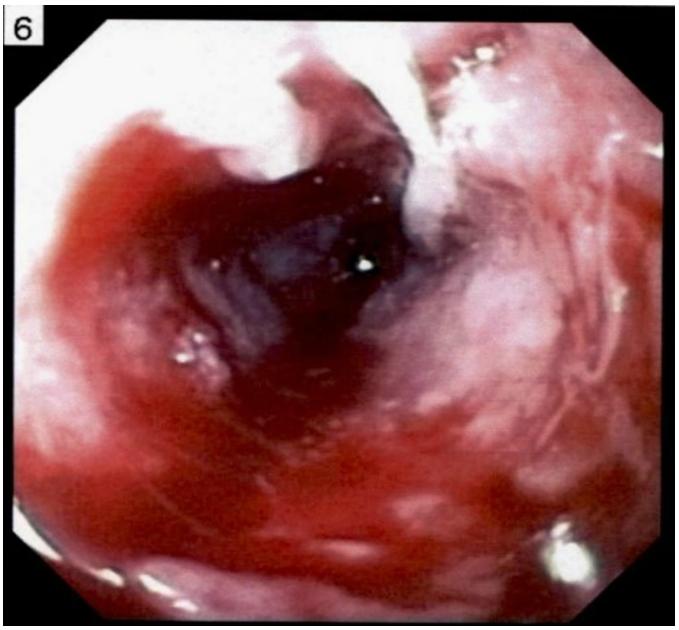

Figure 1: Friable mucosa and pinpoint narrowing of the distal esophagus on initial EGD.

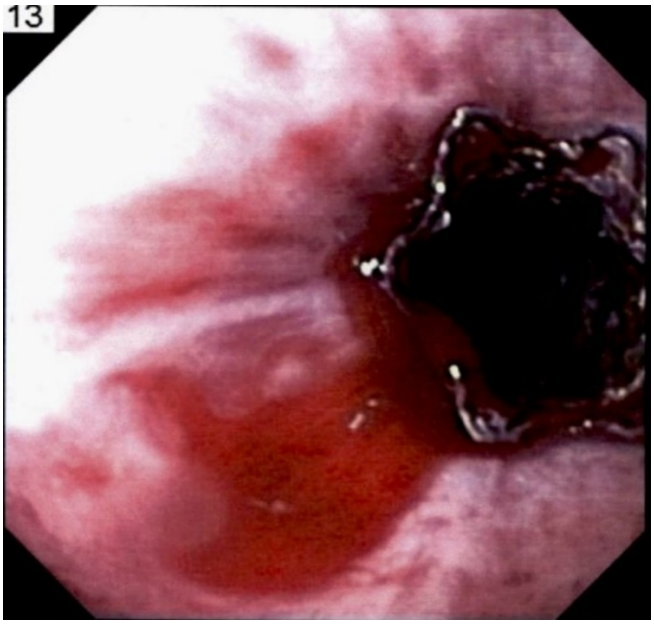

Figure 2: Status-post esophageal stent placement

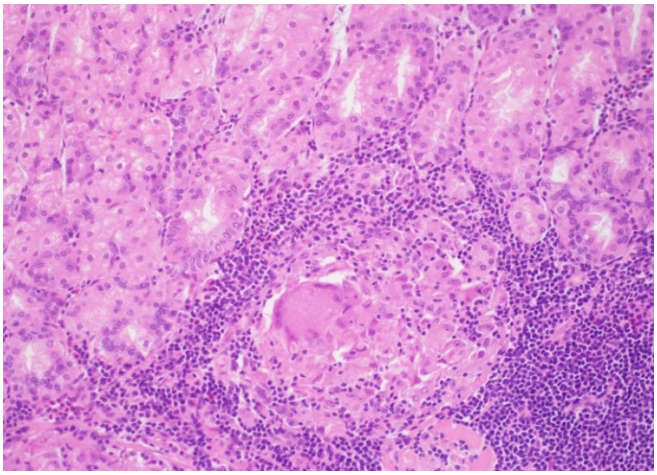
Figure 3: Pathology from the terminal gastric tube biopsy demonstrating noncaseating granuloma

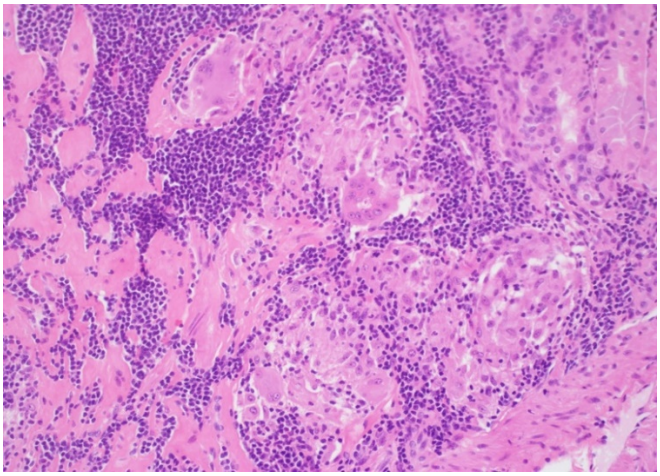
Figure 4: Pathology from the terminal gastric tube biopsy demonstrating noncaseating granuloma

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