CLINICAL REPORT

Sarcoidosis Presenting as Dysphagia

Michael J. Albertson, M.D.

Sarcoidosis is a systemic, chronic, granulomatous disease of unknown etiology that can affect almost any organ system. Gastrointestinal involvement is uncommon, and esophageal involvement is extremely rare. Clinically significant and identifiable gastrointestinal involvement with sarcoid occurs in 0.1-0.9% of patients with systemic sarcoidosis. Case reports of esophageal involvement have been described in the medical and surgical literature.[1,2]

Case Presentation

The patient is a 74-year-old African-American woman referred for persistent dysphagia of solids. She has trouble swallowing pills and has the sensation of food sticking in the lower substernal area. Prior surgical procedures included a hysterectomy with complications of enterotomy and subsequent peritoneal abscess resulting in a left colon resection and colostomy. Because of dense adhesions, the colostomy could not be taken down. A chest X-ray revealed some mediastinal abnormalities, and a CT scan of the chest revealed multicompartmental lymphadenopathy involving the paratracheal, subcarinal, and periesophageal lymph nodes. She was noted to have a subtle reticulonodular pattern of parenchymal disease. A transbronchial biopsy demonstrated a single non-caseating granuloma consistent with sarcoidosis. Upper gastrointestinal endoscopy revealed a tight upper esophageal sphincter, a small greater curvature erosion and no lesions at the lower esophageal sphincter. Biopsies of the erosion revealed nonspecific inflammation. Esophagram demonstrated a fixed and physiologic narrowing at the gastroesophageal junction with a holdup of a swallowed barium tablet. A repeat CT scan of the chest showed significant mediastinal, subcarinal, and perihilar adenopathy similar to the prior scan. The patient was referred for High Resolution Esophageal Impedence Manometry. The resting UES pressure was normal at 49.4 mm Hg. Resting LES pressure was elevated at 77.7 mm Hg (normal 4.8-32 mm Hg). Integrated relaxation pressure was elevated at 31.8 mm Hg (normal < 15 mm Hg). Clearance of a liquid bolus was borderline at 80%. Clearance of a viscous bolus was normal at 90%. The findings were felt to be consistent with outlet obstruction due to functional or anatomical obstruction at the EG junction. Possibilities included stricture, tumor, eosinophilic esophagitis, paraesophageal hernia, lap band, fundoplication, or variant achalasia.

Discussion

Sarcoidosis has the highest incidence in the United States and in Sweden. In the United States, it is more common in African Americans with an annual incidence of 35.5 per 100,000.[3] It occurs more often in women than men. Gastrointestinal involvement in sarcoidosis is very rare. Autopsy studies reported an incidence of subclinical gastrointestinal sarcoidosis of 5-10%. The stomach is the most frequently involved with more than sixty published cases of gastric sarcoidosis.[4] Symptoms include epigastric pain, nausea and vomiting, early satiety, and hematemesis. Gastric sarcoidosis was first described in 1936. Manifestations of sarcoid usually involve ulceration of the gastric mucosa or pyloric lumen by granulomatous infiltration. Pain is present in 75% of these patients. Most cases, however, are subclinical. Other presentations include ulcerative gastric sarcoidosis, infiltrative gastric sarcoidosis and, rarely, polypoid gastric sarcoidosis.[5]

Small intestinal sarcoidosis is the least common form of gastrointestinal sarcoidosis and may present with chronic diarrhea, nausea, abdominal pain, and occasionally malabsorption. Large bowel sarcoidosis can present with a related proctitis or stricture, usually in the sigmoid.

The exact cause of sarcoidosisis is unknown but has been postulated to be due to environmental or infectious etiologies. The characteristic finding in sarcoidosis is the presence of noncaseating granulomata. The two most commonly implicated organisms are mycobacterium and propionobacterium. Sarcoid granulomas contain mycobacterial nucleic acids, and propionibacterial DNA has been found in 98.15% of sarcoidosis lymph node samples.[6]

Esophageal involvement is rare in sarcoidosis with approximately 23 cases reported in the literature.[3] The first case of esophageal sarcoidosis was described by Kerley in 1948 in a patient who presented with dysphagia. An esophagram revealed a distal esophageal stenosis.[7] Of the 23 cases, 91% presented with dysphagia. The lower esophagus (56%) is more commonly involved than the upper esophagus (26%). Dysphagia can occur from direct esophageal wall infiltration, extrinsic compression, cranial neuropathy, and brainstem involvement.[8] Other manifestations are weight loss (22%), abdominal or chest pain (9%), and odynophagia (4%). Heartburn was not reported as an associated in the 23 reported patients. Sarcoidosis can affect the esophagus at different levels. Myopathy involving the skeletal muscle portion of the esophagus and posterior pharynx has been described. A case of Achalasia of the cardia associated with pulmonary sarcoidosis was described in 1983 by Dufresne et al.[4] Because of severe dysphagia a cardiomyotomy was performed. Microscopic diagnosis revealed lesions of the nerves in Auerbach's plexus consisting of an inflammatory process and

demyelinization of the nerve fibers. In 2011, a paper by Bredenoord et al[9] described an Achalasia like dysmotility due to esophageal involvement by sarcoidosis. High resolution manometry revealed absent peristalsis in the esophageal body and incomplete relaxation of the lower esophageal sphincter. The patient's symptoms improved dramatically with treatment with Prednisolone.[9]

The endoscopic appearance of direct esophageal sarcoidosis is nonspecific and includes gray, plaque-like lesions 3-10 mm in diameter, mucosal hyperemia, and nodularity. Biopsies should show non-caseating granulomata with negative special stains for mycobacteria and fungi. This is in addition to the presence of systemic sarcoidosis

Symptoms of dysphagia can be responsive to steroid therapy with Prednisone or Prednisolone. In this case, the patient elected not to take steroids and is doing relatively well with soft foods and nutritional supplements.

REFERENCES

1. **Lukens FJ, Machicao VI, Woodward TA, DeVault KR**. Esophageal sarcoidosis: an unusual diagnosis. J Clin Gastroenterol. 2002 Jan;34(1):54-6. PubMed PMID:11743246.
2. **Wiesner PJ, Kleinman MS, Condemi JJ, Resnicoff SA, Schwartz SI**. Sarcoidosis of the esophagus. Am J Dig Dis. 1971 Oct;16(10):943-51. PubMed PMID: 5120535.
3. **Gallagher P, Harris M, Turnbull FW, Turner L**. Gastric sarcoidosis. J R Soc Med. 1984 Oct;77(10):837-9. PubMed PMID: 6492042; PubMed Central PMCID:PMC1440248.
4. **Dufresne CR, Jeyasingham K, Baker RR**. Achalasia of the cardia associated with pulmonary sarcoidosis. Surgery. 1983 Jul;94(1):32-5. PubMed PMID: 6857509.
5. **Vahid B, Spodik M, Braun KN, Ghazi LJ, Esmaili A**. Sarcoidosis of gastrointestinal tract: a rare disease. Dig Dis Sci. 2007 Dec;52(12):3316-20. Epub 2007 Apr 5. Review. PubMed PMID: 17410465.
6. **Abraham A, Hajar R, Virdi R, Singh J, Mustacchia P**. Esophageal sarcoidosis: a review of cases and an update. ISRN Gastroenterol. 2013 (2013), p. 836203.
7. **Kerley P**. Sarcoidosis. In: Modern Trends in Diagnostic Radiology, McLaren, JW (ed), Hoeber Medical Division, Harper and Row: New York, 1948
8. **Cook DM, Dines DE, Dycus DS**. Sarcoidosis: report of a case presenting as dysphagia. Chest. 1970 Jan;57(1):84-6. PubMed PMID: 5410435.
9. **Bredenoord AJ, Jafari J, Kadri S, Simcock DE, Sifrim D, Preston SL.** Case report: achalasia-like dysmotility secondary to oesophageal involvement of sarcoidosis. Gut. 2011 Feb;60(2):153-5. doi: 10.1136/gut.2010.227868. Epub 2010 Nov 4. PubMed PMID: 21051451.

Submitted September 20, 2015