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Esophageal sarcoidosis presenting as pseudodiverticulum

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Abstract. Sarcoidosis is a multisystem granulomatous disorder that may involve many organs. However, the involvement of the gastrointestinal tract is very rare. This report describes an unusual case of esophageal sarcoidosis presenting as a pseudodiverticulum and reviews the world literature. Our case is also characterized by unusual progression of the esophageal involvement despite stable disease in other organs involved. Myotomy improved the pharyngoesophageal stenosis with no recurrence to date. Physicians should be aware of this rare manifestation, which should be suspected in any sarcoidosis patient complaining about dysphagia. *(Sarcoidosis Vasc Diffuse Lung Dis 2008; 25: 64-67)*

Key words: dysphagia, non-caseating epithelioid granuloma, Zenker diverticulum

Abbreviation List

ACE	Angiotensin-converting enzyme
BALF	Bronchoalveolar lavage fluid
CT	Computed tomography

Introduction

Sarcoidosis is a chronic granulomatous multiorgan disease with unknown etiology. Gastrointestinal tract involvement is very rare, occurring in only 0.1-0.9% of patients (1). Here, we present a case of sarcoidosis involving the upper esophagus.

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Case Report

A 57-yr-old male consulted our hospital with a 18-month history of blurred vision of the right eye in June 1999. He had been previously diagnosed as having uveitis. Chest radiograph showed mild bilateral hilar lymphadenopathy and mild reticulonodular shadows (Fig. 1). Bronchoalveolar lavage fluid

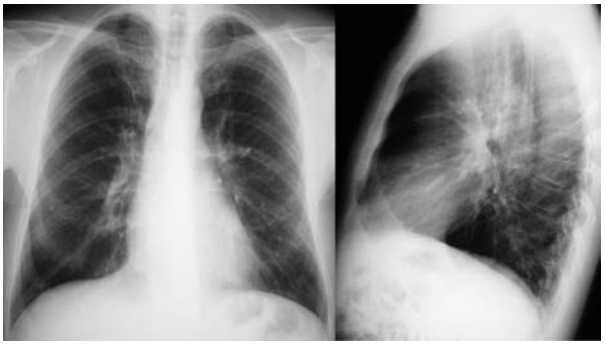

Fig. 1. Chest radiograph showing mild bilateral hilar lymphadenopathy and ill-defined reticulonodular shadows.

(BALF) analysis revealed lymphocytosis (67%) and an increased CD4/CD8 ratio of 3.8 (upper limit of normal 3.5). Serum angiotensin-converting enzyme (ACE) activity was increased to 64 IU/L (normal < 40 IU/L). Pulmonary function tests and blood gas analysis were normal. The patient was asymptomatic. On the basis of these findings, he was clinically diagnosed as having pulmonary sarcoidosis (stage II) complicated with ocular involvement. Systemic steroid therapy (prednisolone 20 mg daily) was administered for ocular involvement. Because of the subsequent stable clinical course as evaluated by ophthalmoscopy, chest radiographs, pulmonary function tests and blood gas analyses, prednisolone was gradually tapered and stopped in 2003.

In March 2004, however, he became symptomatic with dysphagia when eating solid food. Chest radiographs, pulmonary function tests and blood gas analyses revealed no change. Chest computed tomography (CT) demonstrated enlarged pretracheal lymph nodes without tracheal compression, unchanged with the previous CT examinations. There was no adenopathy in the area of the upper esophagus. No pharyngeal involvement was seen on CT. In esophagography and a swallowing study, no complications were detected except mild gastroesophageal reflux revealed by esophageal pH monitoring. Serum ACE activity was slightly increased to 89 IU/L.

In June 2006, the reexamined esophagography suggested the possibility of a Zenker diverticulum in the upper esophagus (Fig. 2). Left-lateral cervicotomy with complete exploration of the hypopharynx and upper esophagus, however, revealed no signs of a cervical diverticulum. A complete lateral myotomy of the hypopharynx and proximal esophagus with a biopsy of the esophageal wall was performed. Histopathological examination showed non-caseating epithelioid granuloma in the wall of the esophagus (Fig. 3). Subsequently, the dysphagia for solid food improved. To date, the patient remained asymptomatic without the need for systemic steroid therapy.

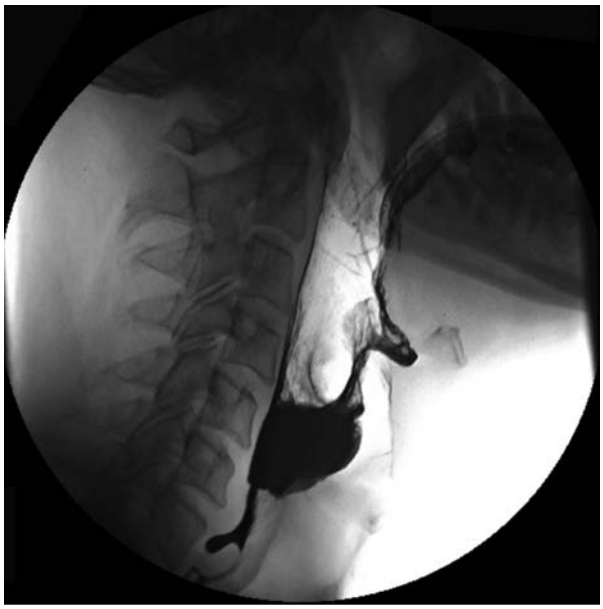

Fig. 2. Esophagography showing a diverticulum at the posterior wall of the upper esophagus. No esophageal stenosis is observed.

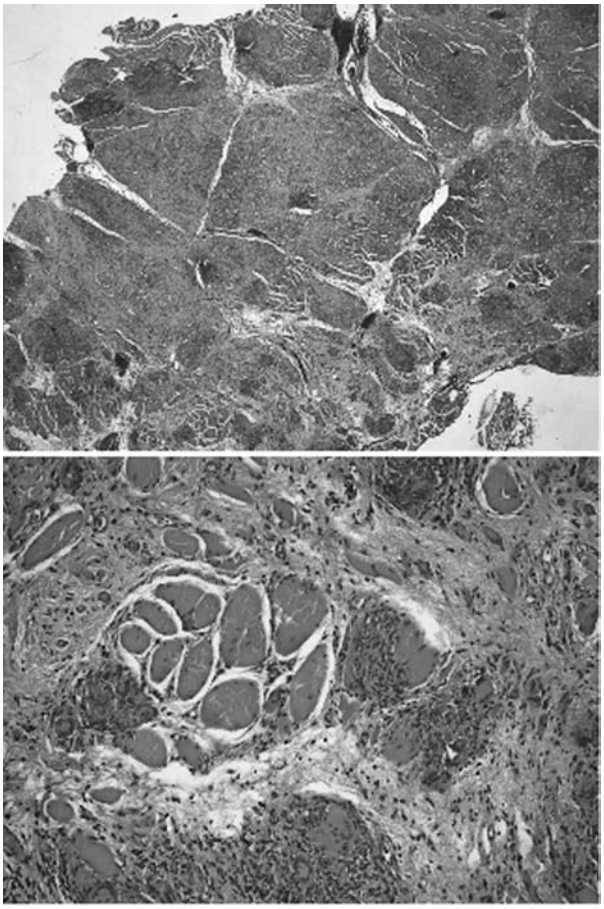

Fig. 3. Hematoxylin-eosin stain of the wall of the esophagus showing an infiltration consisting of multiple epithelioid cell granulomas with giant cells and some lymphocytes (20 x) (*top*), at higher magnification, classical non-caseating epithelioid granuloma with giant cells, infiltrating smooth muscle bundles (400 x) (*bottom*).

Table 1. 15 cases with esophageal sarcoidosis (Review of world literature and own case).

Author	Year	Involvement	Symptoms	Cause
Siegel CI (9)	1961	Pharyngoesophageal junction	Dysphagia	Direct granulomatous infiltration
Panosetti E (10)	1979	Pharyngoesophageal junction	Dysphagia	Direct granulomatous infiltration
Ohshimo S	2008	Pharyngoesophageal junction	Dysphagia	Direct granulomatous infiltration
Davies RJ (11)	1972	Upper esophagus	Dysphagia	Direct granulomatous infiltration
Cook DM (2)	1970	Mid esophagus	Dysphagia	Extrinsic compression by enlarged lymph nodes
Kerly P (12)	1948	Lower esophagus	Dysphagia	Direct granulomatous infiltration
Polachek AA (5)	1964	Lower esophagus	Abdominal pain	Direct granulomatous infiltration
Hardy WE (6)	1967	Lower esophagus	Dysphagia, Dysphonia	Direct granulomatous infiltration
Wiesner PJ (3)	1971	Lower esophagus	Dysphagia	Direct granulomatous infiltration
Lukens FJ (4)	2002	Lower esophagus	Dysphagia, Achalasia	Direct granulomatous infiltration
Murdock A (1)	2003	Lower esophagus	Anemia, Barrett's esophagitis	Direct granulomatous infiltration
Dufresne CR (13)	1983	Lower esophagus	Dysphagia, Achalasia	Neural invasion with granulomas
Aronson PJ (14)	1985	Lower esophagus	Dysphagia, Achalasia, Hoarseness	Neural invasion with granulomas
Nidiry JJ (7)	1991	Lower esophagus	Dysphagia, Achalasia	Neural invasion with granulomas
Boruchowicz A (8)	1996	Lower esophagus	Dysphagia, Achalasia	Neural invasion with granulomas

Discussion

Here we report a case of esophageal sarcoidosis presenting with dysphagia, a pseudodiverticulum and severe pharyngoesophageal stenosis. Histopathology showed infiltration of the smooth muscle of the esophageal wall by sarcoid granulomas. There was no evidence of indirect compression of the esophagus by enlarged mediastinal lymph nodes.

Previous reports have presented various pathogenetic pathways for dysphagia in patients with sarcoidosis; 1) compression of the esophagus from outside the walls by enlarged lymph nodes (2), 2) achalasia associated with a contractile disorder of the cardia derived from neural invasion with granulomas and 3) direct infiltration of the esophageal wall by granulomas (3). Esophagram, esophageal motility study and histopathological examination of the tissue obtained by esophagogastroscopy, thoracoscopy, mediastinoscopy or thoracotomy have been reported to be beneficial for the diagnosis of esophageal sarcoidosis (1, 4). However, the upper esophageal regions may frequently show false-negative findings in the esophagram or esophagogastroscopy due to technical limitations, as in the first esophagram of our case. Accordingly, the investigation should predominantly be focused on the entrance and the upper part of the esophagus in sarcoidosis patients with prolonged dysphagia. Zenker diverticulum is an acquired compressive diverticulum that is formed at the posterior wall of the upper esophagus resulting from the permanent increment of intraesophageal pressure. In our case, the lesion was initially regarded as Zenker diverticulum, but was possibly a pseudodiverticulum caused by the stenosis of the upper esophagus.

A remarkable finding in the present case is the discrepancy between the progression of pharyngoesophageal involvement and the stabilized pulmonary state as evaluated by physiological and radiological examinations. In the present case, no examination before the operation was able to demonstrate the pharyngoesophageal involvement.

Because of the very limited number of cases with esophageal sarcoidosis (Tab. 1), no standard therapy has been established. Previous studies have reported the benefits of oral steroid therapy (2, 5-7), distal esophageal myotomy (8), cricopharyngeal myotomy (9) and local botulinum toxin injection (4) in patients presenting with dysphagia. In the present case, myotomy improved the dysphagia and maintained the asymptomatic state to date.

In summary, we have presented a case of pharyngoesophageal sarcoidosis demonstrating a pseudodiverticulum and pharyngoesophageal stenosis. Physicians should be aware of the possibility of this rare manifestation as cause of dysphagia in a patient with sarcoidosis.

References

1. Murdock A, Jacob G. Sarcoidosis of the esophagus presenting macroscopically as Barrett's esophagitis. Am J Gastroenterol 2003; 98: 1661-2.
2. Cook DM, Dines DE, Dycus DS. Sarcoidosis: report of a case presenting as dysphagia. Chest 1970; 57: 84-6.

3. Wiesner PJ, Kleinman MS, Condemi JJ, et al. Sarcoidosis of the esophagus. Am J Dig Dis 1971; 16: 943-51.
4. Lukens FJ, Machicao VI, Woodward TA, et al. Esophageal sarcoidosis: an unusual diagnosis. J Clin Gastroenterol 2002; 34: 54-6.
5. Polachek AA, Matre WJ. Gastrointestinal Sarcoidosis. Report of a Case Involving the Esophagus. Am J Dig Dis 1964; 9: 429-33.
6. Hardy WE, Tulgan H, Haidak G, et al. Sarcoidosis: a case presenting with dysphagia and dysphonia. Ann Intern Med 1967; 66: 353-7.
7. Nidiry JJ, Mines S, Hackney R, et al. Sarcoidosis: a unique presentation of dysphagia, myopathy, and photophobia. Am J Gastroenterol 1991; 86: 1679-82.
8. Boruchowicz A, Canva-Delcambre V, Guillemot F, et al. Sarcoidosis and achalasia: a fortuitous association? Am J Gastroenterol 1996; 91: 413-4.
9. Siegel CI, Honda M, Salik J, et al. Dysphagia due to granulomatous myositis of the cricopharyngeus muscle; physiological and cineradiographic studies prior to and following successful surgical therapy. Trans Assoc Am Physicians 1961; 74: 342-52.
10. Panosetti E, Lehmann W. [Localized sarcoidosis of the cervical oesophagus (author's transl)]. Schweiz Rundsch Med Prax 1979; 68: 349-53.
11. Davies RJ. Dysphagia, abdominal pain, and sarcoid granulomata. Br Med J 1972; 3: 564-5.
12. Kerly P. Sarcoidosis. In: McLaren JW, ed. Modern trends in diagnostic radiology. New York: Hoeber Medicine Division: Harper & Row 1948: 150-2.
13. Dufresne CR, Jeyasingham K, Baker RR. Achalasia of the cardia associated with pulmonary sarcoidosis. Surgery 1983; 94: 32-5.
14. Aronson PJ, Fretzin DF, Morgan NE. A unique case of sarcoidosis with coexistent collagen vascular disease. Possible result of a compatible disease-sustaining immunologic environment. J Am Acad Dermatol 1985; 13: 886-91.