Mediastinal tuberculous lymphadenitis presenting as an esophageal intramural tumor: A very rare but important cause for dysphagia

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Abstract

Dysphagia associated with esophageal mechanical obstruction is usually related to malignant esophageal diseases. Benign lesions are rarely a cause for this type of dysphagia, and usually occur either as an intramural tumor or as an extrinsic compression. Mediastinal tuberculous lymphadenitis is rare in adults, and even more rarely causes dysphagia. We report two cases of dysphagia in adult patients caused by mediastinal tuberculous lymphadenitis, presenting radiologically and endoscopically as an esophageal submucosal tumor. Based on the clinical and imaging diagnosis, the patients underwent a right thoracotomy, and excision of the mass attached to and compressing the esophagus. Pathological examination of the specimens showed a chronic granulomatous inflammation with caseous necrosis, which was consistent with tuberculous lymphadenitis.

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Key words: Dysphagia; Tuberculous lymphadenitis; Esophageal tumor; Uncommon dysphagia; Esophageal benign lesion

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INTRODUCTION

In the esophagus, squamous cell adenocarcinoma as the most common type of malignant tumors and leiomyoma as the most common type of mesenchymal neoplasms, represent the frequent causes for dysphagia. However, dysphagia associated with esophageal mechanical obstruction can be caused by several conditions, mainly endoluminal/mucosal lesions, intramural tumors or extrinsic compression.

Despite technological improvements in the diagnostic work-up, the definitive diagnosis of some of these entities, particularly differentiation of submucosal parietal lesions from extrinsic lesions compressing the esophagus, may only be established after surgery[1].

Mediastinal diseases such as sarcoidosis[2], lung cancer, lymphoma, distant metastases[3-6] or mediastinal inflammatory lymph nodes[1,5-9] are the conditions that rarely compress the thoracic esophagus and cause dysphagia.

Some causes for dysphagia should always be considered in the differential diagnosis of dysphagia, because some of them if diagnosed and treated in time will completely cure the patient. We herein report two cases of dysphagia caused by mediastinal tuberculous lymphadenitis (MTL), presenting as esophageal intramural tumors.

CASE REPORT

Case 1

A 40-year-old woman who complained of a six-month history of dysphagia for solid food and paroxystic odynophagia, was admitted to our hospital for further evaluation of a submucosal mass-like lesion seen on an upper gastrointestinal (GI) endoscopy taken in another hospital. She had no other digestive symptoms, denied weight loss, fever, cough, sputum and night sweating. Her past medical history was unremarkable, namely exposure to tuberculosis and her family history was also unremarkable. Clinical examination was irrelevant, and her hematological and biochemical tests were normal.

A chest X-ray did not show any parenchymal and pleural change or mediastinal pathology. A barium swallow esophagogram (Figure 1A) disclosed a two-centimeter protruding lesion on the left lateral aspect of the mid-esophagus, distal to the aortic arch and with a smooth surface. An upper GI endoscopy (Figure 1B) showed a
two-centimeter deformity of the esophageal wall, covered by normal mucosa and with firm consistency, located 25 centimeters from the anterior incisor teeth. An endoscopic ultrasonography (Figure 1C) suggested that the lesion described by the other methods was an extramucosal mesenchymal tumor of the esophagus. A CT scan showed no pulmonary lesions or enlargement of mediastinal lymph nodes. The patient underwent a right thoracotomy, and excision of the mass attached to and compressing the esophagus. Surgery was performed with some difficulty due to surrounding fibrosis.

Pathological examination of the specimen showed a conglomerate of lymph nodes surrounded by chronic granulomatous inflammation with caseous necrosis. Zielh-Neelsen stain of the specimen, however, failed to disclose acid-fast bacilli (Figure 1D).

The symptoms of dysphagia and odynophagia disappeared after excision of the mass and postoperative treatment with a three-regimen anti-tuberculous chemotherapy with isoniazid, ethambutol hydrochloride and rifampicin.

A follow-up barium swallow esophagogram revealed a normal esophagus and the patient was in good health 6 years after an uneventful operation and subsequent discharge.

Case 2
A 54-year-old woman was referred to our hospital with a five-month history of dysphagia for solid food and a diagnosis of a submucosal lesion in the middle third of the esophagus. She had no other digestive symptoms, denied weight loss, fever, cough, sputum and night sweating. Her past medical history was unremarkable, namely exposure to tuberculosis and her family history was also unremarkable. Clinical examination was irrelevant, except for a palpable right supra-clavicular lymph-node. In spite of a repeatedly high erythrocyte sedimentation rate (30-73 mm in the first hour), sputum examination to disclose acid-fast bacilli and all the other routine laboratory tests revealed no abnormal findings.

Fine needle cytology of the supra-clavicular lymphadenopathy showed a necrotic lesion, without malignant cells. Zielh-Neelsen stain was negative. A barium swallow esophagogram (Figure 2A and B) disclosed a four-centimeter protruding smooth lesion on the right lateral aspect of the mid-esophagus, with apparent integrity of mucosa. An upper gastrointestinal (GI) endoscopy (Figure 2C) showed a two-centimeter deformity of the esophageal wall covered by normal mucosa and with firm consistency, located 22 centimeters from the anterior incisor teeth. An endoscopic ultrasonography (Figure 2D) revealed a two-centimeter hypoecogenic mass with well-defined limits, which was apparently dependent on the fourth ecographic layer of the esophagus, suggesting a leiomyoma. A chest X-ray and a CT scan did not show any lung parenchymal or pleural change, but the CT scan revealed enlargement of some mediastinum and pulmonary hilum lymph nodes, even though they did not form conglomerates.

Through a right thoracotomy, the mass compressing the esophagus was removed, in fact, an enlarged lymph node, and some other lymph nodes adherent to the esophageal wall were also removed.

Pathological examination of the specimen showed lymph nodes surrounded by a chronic granulomatous inflammation with caseous necrosis. Zielh-Neelsen stain of the specimen disclosed acid-fast bacilli (Figure 2E).

The patient began a postoperative three-regimen anti-tuberculous treatment with isoniazid, ethambutol hydrochloride and rifampicin.

The symptoms of dysphagia disappeared after treatment and a follow-up barium swallow esophagogram (Figure 2F) revealed a normal esophagus. The patient was in good health 4 years after discharge.

DISCUSSION
The ethiology of dysphagia associated with esophageal mechanical obstruction is usually referred to as
endoluminal/mucosal, intramural or extrinsic, in accordance with the location of the initial lesion in the esophageal wall. Malignant tumors of the esophagus are the most frequent cause, not only for primary dysphagia, but also for dysphagia caused by lesions originating from the esophageal mucosa\[10\].

Obstructive dysphagia caused by benign lesions is rare and usually results from intramural or extrinsic lesions. Even though rare, benign lesions leading to dysphagia are of great concern in the clinic. In fact, these lesions, if not treated, can lead to death due to progressive obstruction of the esophagus or pulmonary complications, but they almost always allow a curative surgical resection\[10\].

The endoscopic and radiological characteristics of a submucosa tumor are the endoluminal protrusion of the digestive tube wall, usually covered by a normal looking mucosa. However, such features may also be caused by organ lesions or structures that extrinsically compress the digestive wall. EUS is described as the diagnostic method with a greater capacity of distinguishing between intraparietal and extrinsic lesions compressing the digestive wall, with a diagnostic accuracy superior to CT-scan and barium swallow\[11,12\]. Although EUS-guided fine needle biopsy is possible in these lesions, the material retrieved is sometimes insufficient for a diagnosis and, particularly for evaluation of the malignant potential of the lesions\[12-15\].

In our cases, even though the diagnosis of an extrinsic compression on the esophagus could not be excluded, barium swallow revealed an image compatible with a submucosa tumor protruding into the lumen. In fact, the angle between the esophageal wall and margin of the mass was almost perpendicular, which is in accordance with an intramural lesion, rather than an extrinsic compression on the esophagus\[16\]. Furthermore, all of the other diagnostic examinations pointed towards the diagnosis of a submucosal tumor-like intraluminal protruding mass. For these reasons, the patients were operated on with a probable diagnosis of a leiomyoma of the esophagus, suggesting that this is the most frequent intraparital benign tumor of the esophagus.

One of our patients had a supraclavicular adenopathy and a persistently high erythrocyte sedimentation rate. The lymph node biopsy revealed necrotic material, but no Zielh-Neelson stained bacilli. Even though central necrosis of a lymph node is a frequent finding in tuberculous lymphadenitis\[17\], this fact does not give us a diagnosis of tuberculosis. In our cases, pulmonary Rx and CT scan of the thorax did not reveal lesions compatible with the diagnosis of pulmonary tuberculosis.

Mediastinal inflammatory lymph nodes, particularly tuberculous lymphadenitis\[16,18-20\] is a condition compressing the thoracic esophagus and causing dysphagia. Although mediastinal tuberculous lymphadenitis is rare, it is increasing in adults\[18-20\]. On the other hand, tuberculosis is an infectious disease with a rising incidence particularly in Asian and Eastern European countries and also a rising prevalence in association with HIV infection\[21\]. In 1993, tuberculosis was declared as a global emergency, by the World Health Organization\[22\].

Mediastinal tuberculous lymphadenitis should be included in the differential diagnosis of dysphagia, but it should be realized that it can present with various endoscopic and radiological findings. In fact, mediastinal tuberculous lymphadenitis can affect the esophagus by compressing the esophagus externally, causing rupture in the mediastinum and leading to an inflammatory process with secondary involvement of the esophagus, invading...
the esophagus, ulcerating the mucosa and draining caseum into the esophageal lumen\cite{2}, as well as resulting in an esophageal fistula in some cases\cite{21,22}.

Dysphagia in mediastinal tuberculous lymphadenitis is due to the external compression on the esophagus, but the pain during swallowing - odynophagia occurring in one of our patients, suggested that the esophagus is directly involved in the inflammatory process. Interestingly, Ghimire and Walker\cite{1} reported a case of mediastinal tuberculous lymphadenopathy who had painful dysphagia without significant involvement of the esophagus endoscopically and radiologically. They assumed that the contiguous inflammation of paraesophageal tissues resulted in disturbed esophageal motility of the patient.

In cases of tuberculous adenitis with esophageal mucosal lesion, confirmation of diagnosis should be done by histological or microbiological examination of the specimen obtained by endoscopic biopsy. In our cases without lesion of the mucosa, but with a strong suspicion that the cause for dysphagia is of tuberculous origin, EUS and endoscopic fine needle biopsy can play a role in diagnosing mediastinal masses that produce esophageal symptoms\cite{4,11}. Other authors prefer biopsies guided by mediastinoscopy or thoracoscopic, when there is no mucosal lesion\cite{8,20}. Nevertheless, when it is impossible to reach the affected lymph nodes through these approaches, surgery may be necessary in order to establish a diagnosis\cite{16}.

At present, minimally invasive surgery should be attempted to remove lesions diagnosed pre-operatively as probable leiomyomas. However, it was difficult, in the first case, to localize the lesion immediately below the aortic arch, on the left side of the esophagus associated with the surrounding fibrosis. In fact, it is hard to isolate the lesion in the presence of mediastinitis fibrosis resulting from rupture of an affected lymph node\cite{21-23}. Medical treatment of most patients with tuberculosis consists of a short course in chemotherapy, using 3 or 4 essential anti-tuberculosis drugs (isoniazid, rifampicin, ethambutol, pyrazinamide and streptomycin). Anti-TB regimen consists of an initial intensive phase and a continuation phase\cite{17-20}. It was reported that most patients can be successfully treated with a three-drug anti-tuberculosis chemotherapy regimen\cite{6,60,23}. If the diagnosis of mediastinal tuberculous lymphadenitis is made pre-operatively, such a treatment may lead to favourable outcome\cite{10,23}. Surgery should be reserved for patients with no pre-operative diagnosis, as in our cases, and those who develop complications, such as mediastinal abscess that is unresponsive to non operative management\cite{21}.

Since the diagnosis of a small esophageal submucosal lesion or an intraparietal lesion is usually difficult, surgery is necessary. Even though rare, the diagnosis of mediastinal tuberculous lymphadenitis should be highly considered in the presence of an uncertain esophageal lesion\cite{23}. This is particularly important because a diagnostic non surgical approach is possible and an adequate treatment can completely cure mediastinal tuberculous lymphadenitis with or without tuberculous esophagitis\cite{10}.

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