Mediastinal Lymphangioma Treated Using Endobronchial Ultrasound-Guided Transbronchial Needle Aspiration

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Established Facts
- The standard treatment of mediastinal lymphangioma has been surgical excision. However, the involvement of vital structures in the area local to the lymphangioma makes total excision virtually impossible in most cases.

Novel Insights
- Endobronchial ultrasound-guided transbronchial needle aspiration may be a safe, effective, new alternative treatment for mediastinal lymphangioma that avoids the unfavorable outcomes associated with other treatments.

Key Words
Mediastinal lymphangioma • Endobronchial ultrasound • Transbronchial needle aspiration

Abstract
Lymphangiomas are localized malformations of the lymphatic system that most commonly occur in the head and neck. However, less than 1% of all lymphangiomas are confined to the mediastinum. The standard treatment has been surgical excision, but the involvement of vital structures in the area local to the lymphangioma makes total excision virtually impossible in most cases. To our knowledge, there has been no report of mediastinal lymphangioma treated with endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA). We report here the first case of safe, effective treatment of a very large mediastinal lymphangioma using EBUS-TBNA in a 29-year-old man.
Introduction

Lymphangiomas are rare benign lesions characterized by the focal proliferation of well-differentiated lymphatic tissue that present as multicystic or sponge-like accumulations. Less than 1% of all lymphangiomas are confined to the mediastinum. Although the majority of lymphangiomas present in the first 2 years of life, recognition of lymphangiomas in adults has recently increased. The treatment options for lymphangioma are percutaneous aspiration, laser therapy, percutaneous sclerotherapy and surgery. However, treatment of mediastinal lymphangioma remains difficult [1–4]. To our knowledge, no mediastinal lymphangioma treated with endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) has been reported in the English literature. This study reports the first successful treatment of a mediastinal lymphangioma in an adult using EBUS-TBNA.

Case Report

A 29-year-old man was admitted to the hospital with a perforated duodenal ulcer. The preoperative chest x-ray and computed tomography (CT) showed a homogeneous, low attenuation, cystic mediastinal mass in the paratracheal area (fig. 1a, b). The mass extended from the inferior aspect of the thyroid gland down to the carina. Its maximum transverse diameter was 13.7 cm. The patient had abdominal pain and an intermittent dry cough, but was well otherwise. His family history was noncontributory. Ten years earlier, the mediastinal mass had been found incidentally during a medical check-up for his military enlistment. However, no treatment was considered in two centers owing to the technical difficulty and the fact that he was asymptomatic. The current CT showed a significant increase in mass size, with the maximum transverse diameter increasing from 9.3 to 13.7 cm compared with the CT of 10 years earlier.

At bronchoscopy, the lower trachea was found to be mildly compressed with no mucosal lesion (fig. 2a). EBUS with a convex probe (BF-UC260F-OL8, Olympus, Tokyo, Japan) was performed to diagnose the mass and to observe an anechoic lesion with no Doppler flow signal (fig. 2b). TBNA was performed using a dedicated 22-gauge needle (fig. 2c). Straw-colored serous fluid was aspirated, and the cytological examination revealed only mature benign lymphoid cells (fig. 2d) with negative microbiological culture, suggestive of a lymphangioma. The patient underwent laparoscopic primary closure of a duodenal ulcer as scheduled. One week later, EBUS-TBNA was selected as the therapeutic technique of choice at a multidisciplinary conference, which included the patient’s opinion. Chest magnetic resonance imaging (MRI) performed prior to treatment showed a multiseptated cystic lesion around the trachea (fig. 3). The EBUS-TBNA was performed under local anesthesia with mild conscious sedation using midazolam. In total, 700 ml were aspirated over 2 days, over 60 and 80 min...
The patient was discharged from hospital on the third day after treatment, with no complications. At 1, 3 and 12 months after treatment the patient was followed up to assess any delayed complications or recurrence. Follow-up chest x-rays and CT after treatment showed a marked decrease in the volume of the mediastinal lymphangioma (fig. 1c, d), with no delayed complications.

**Discussion**

Mediastinal lymphangiomas are rare and constitute approximately 0.01–4.5% of all mediastinal masses [4]. Most mediastinal lymphangiomas are asymptomatic unless they grow enough to press on adjacent structures. Ultimately, the diagnosis is made after a histological examination of resected tissue. However, the presence of benign lymphoid aggregates, such as in our case, is helpful in the identification of lymphangiomas. MRI is probably the diagnostic modality of choice for lymphangiomas, because it accurately predicts the subsequent intraoperative findings, and it helps to demonstrate the lymphatic architecture at different tissue levels [2]. Generally, classification of mediastinal cysts is based on their etiology, encompassing bronchogenic, esophageal duplication cysts of foregut origin, mesothelial derived pericardial or pleural cysts, thymic cysts and other miscella-

**Fig. 2.** a Bronchoscopic image showing mildly extrinsic compression of the lower trachea. b, c EBUS images of the right lower paratracheal lesion showing a homogeneous anechoic lesion with no Doppler flow signal and the needle inserted in the anechoic lesion. d Aspirated smear of the mediastinal lymphangioma showing numerous mature benign lymphoid cells (HE staining. ×400). e A photograph of the straw-colored fluid aspirated from the patient’s mediastinal lymphangioma via EBUS-TBNA.

**Fig. 3.** T2-weighted coronal MRI performed prior to treatment showing multiple internal septation with high signal intensity.
Although lymphangiomas are benign lesions, some treatment is necessary because of the problems they may cause. With lymphangiomas in the mediastinum and neck region, acute complications such as airway obstruction and infection may arise. In general, the standard treatment has been surgical excision. Complications from surgical excision have been reported in 10–33% of cases. Intraoperative blood loss, muscle weakness, Horner’s syndrome, nerve paralysis, recurrent airway obstruction, life-threatening infections such as mediastinitis and sepsis, chylothorax and death are some of the more serious postoperative complications. However, the involvement of vital structures in the lymphangioma area makes total excision almost impossible in most cases. Furthermore, recurrence rates range from 0 to 27% and 50 to 100% following complete or incomplete excision, respectively [2–4]. Although surgery remains an excellent therapeutic technique for microcystic and mixed lesions, some authors now recommend sclerotherapy as the primary therapy for macrocystic lesions. Hence, the treatment of lymphangiomas is challenging. Although the current level of supporting evidence is only fair, sclerotherapy has been effective for some lymphangiomas [3, 6, 7].

Conventional TBNA may have utility in other clinical situations, such as in the diagnosis and treatment of primary mediastinal cysts or even of pericardial effusions [8, 9]. However, recent progress with EBUS technology has made it easier to obtain cytological and histological samples from lesions adjacent to the tracheobronchial tree [10–12]. Convex-probe EBUS also enables real-time procedures to be performed under ultrasound guidance, and allows deep, complete aspiration, resulting in the collapse of the mediastinal cystic lesions [12]. In our case, surgery was considered initially, but complete resection of the mediastinal lymphangioma was felt to be too difficult. Therefore, EBUS was selected as a new alternative treatment technique. In patients with mediastinal lymphangioma, EBUS-TBNA appears to be a safe, effective treatment option. Furthermore, the EBUS system should facilitate the safe injection of sclerosing agents using a needle assembly system for EBUS-TBNA after aspiration of the cystic fluid under ultrasound guidance. In our case, we considered EBUS-TBNA only as an initial and minimally invasive treatment for a slow-growing (about 47% increase in the long diameter in 10 years) and incidentally recognized mediastinal lymphangioma. However, we will consider additional sclerotherapy using the EBUS system if the mediastinal lymphangioma is again large in mass during follow-up.

In conclusion, EBUS-TBNA may be a safe, effective, new alternative treatment for mediastinal lymphangioma that avoids the unfavorable outcomes associated with other treatments.

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References