Primary amyloidosis involving mediastinal lymph nodes diagnosed by EBUS-TBNA

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Summary
Amyloidosis is an uncommon disease caused by the deposition of abnormal proteins within the soft tissues. Mediastinal lymph node involvement of the disease is rare. When mediastinal lymph nodes are affected, tissue sampling is required for obtaining a firm diagnosis. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) is a minimally invasive method of sampling tissues from mediastinal and hilar lymph nodes. We report a case of primary amyloidosis involving mediastinal lymph nodes successfully diagnosed by EBUS-TBNA.

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Introduction
Amyloidosis is an uncommon disorder resulting from the deposition of a fibrillar protein in tissues. 1 Lymph node amyloid deposits lead to lymph node enlargement. 2 Diagnosis is usually difficult and made through thoracotomy or mediastinoscopy. 3 But, it is invasive, requires general anesthesia and complication cannot be ignored. 4 Direct real-time endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) is a minimal invasive modality for tissue sampling of the mediastinum. 5,6 This is the first reported case of intrathoracic amyloidosis affecting mediastinal lymph nodes diagnosed by histological assessment of tissue obtained by EBUS-TBNA.

Case report
A 77-year-old woman was referred to our department who presented with cough and hoarseness with bilateral hilar and mediastinal lymphadenopathy on chest computed tomography (CT) in 2006. She had been followed as suspected sarcoidosis with bilateral hilar lymphadenopathy on
annual medical examination from 2004. In 2006, chest CT showed continuous lymphadenopathy from neck to the hilum (Fig. 1A). She also had lower chin lymphadenopathy, as well as swelling of the rhinopharynx. She was suspected as sarcoidosis and percutaneous needle biopsy of the rhinopharynx was performed. It showed no granuloma, but some eosinophil deposition suggested of amyloidosis. However, pathological results were not diagnostic. EBUS-TBNA was performed from the subcarinal lymph node to obtain lymphoid tissue (Fig. 1B). The convex probe endobronchial ultrasound (XBF-UC260F-OL8, Olympus, Tokyo) was used for EBUS-TBNA. Hyaline-like amorphous deposits were seen on cytology and eosinophil depositions were seen which were positive for Congo-Red staining on pathology. Furthermore, polarizing microscope showed a unique yellowish-green polarized space (Fig. 2). The final pathological diagnosis was primary amyloidosis. Serum immuneelectrophoresis disclosed a monoclonal peak of immunoglobulin light chains with elevated total IgM level. Urine Bence-Jones protein assay was positive. β2 microglobulin was slightly elevated by renal examination. Slight dysautonomia and slight lower limbs predominance axon denaturation type neuropathy were seen in the neurological examination. On bone marrow biopsy, only a few plasma cells were seen. Cardiac dysfunction due to deposition of amyloid was not seen. The patient has been followed up without treatment due to her age.

Discussion

EBUS-TBNA allows investigation of the majority of the mediastinum comparable to the reach of the mediastinoscope. Compared to mediastinoscopy, EBUS-TBNA is
minimally invasive, can be done quite safely, and can be performed under local anesthesia in an outpatient setting.5,6 EBUS-TBNA allows cytological as well as histological examination from mediastinal lymph nodes. To obtain a histological sample, the dedicated 22-gauge needle is used equipped with an internal sheath. The internal sheath is used to clean out the internal lumen after initial puncture as well as used to push out the histological core.5,6 By obtaining histological cores, we have successfully diagnosed different types of mediastinal tumors.7

Amyloidosis is a heterogeneous group of disorders characterized by extracellular deposition of abnormal proteins.8 Although the precise etiology of amyloidosis is not known, the abnormal protein deposition may result from a disordered immune response to a prolonged antigenic challenge. Amyloidosis may be localized to one organ or body tissue, but more often it is systemic. The systemic form can be primary, associated with multiple myeloma, secondary to a chronic disease (such as rheumatoid arthritis and Crohn’s disease), malignancies, or familial.2,3 Localized amyloidosis has a good prognosis, but systemic amyloidosis often causes death within several years of diagnosis. Heart disease and renal failure are common causes of death.3 Thoracic manifestations can occur in both the localized and systemic forms of amyloidosis.9

The leading causes of mediastinal lymphadenopathy include infections, sarcoidosis, Castleman’s disease and neoplastic diseases.2,10 On the other hand, mediastinal amyloid lymphadenopathy is extremely rare.2 Furthermore, amyloidosis should be diagnosed based on pathological confirmation.3 Patients with systemic amyloidosis often have cardiac involvement and this ultimately causes death in many patients with this disease.3 Performing surgical biopsy under general anesthesia may be a risk in patients with systemic amyloidosis. Non-surgical biopsies should be considered if there are possibilities of mediastinal involvement.2 EBUS-TBNA may be an alternative to surgical biopsies for the diagnosis of amyloidosis when mediastinal lymphadenopathy is seen.

Conflict of interest Statement

We have no relations to disclose to this article.

References