Primary Laryngo-Tracheobronchial Amyloidosis: A Case Report and Literature Review

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Abstract

Primary laryngo-tracheobronchial amyloidosis is a rare pulmonary disease for which there is currently no established treatment. Herein, we report a case involving a 51-year-old woman who was admitted to hospital primarily due to intermittent hoarseness. Botryoidal masses were visible at the entrance of the throat. On chest computed tomography, eccentric soft tissue density shadows were apparent in the tracheal bifurcation of the proximal segment of the trachea, and the lumen was narrowed. Electronic bronchoscopy revealed extensive new biological invasive growth from the nasopharynx to the glottis. The glottis and larynx were irregularly shaped with clear characteristics and congestion. New biological growth was also observed in the right upper lobe, right middle bronchus, and right lower lobe mucosa. The patient was ultimately diagnosed with primary laryngo-tracheobronchial amyloidosis via pathological analysis. Although amyloidosis is a benign lesion, to date, there are no curative treatments. The present article briefly highlights the pertinent literature, and discusses the clinical manifestations and progress of treatment in primary laryngo-tracheobronchial amyloidosis.

Keywords: Laryngo-tracheobronchial; Amyloidosis; Pulmonary; Treatment

Introduction

Amyloidosis refers to the extracellular deposition of amyloid, a fibrillar proteinaceous insoluble material with characteristic light, ultrastructural, and histochemical features. It consists of a well-circumscribed, dense, amorphous, eosinophilic amyloid protein surrounded by an inflammatory cellular infiltrate of plasma cells and lymphocytes, and is occasionally accompanied by a granulomatous reaction [1]. It is a relatively rare condition with low morbidity (approximately 0.0008% per year) [2]. Amyloidosis often involves the heart, liver, kidney, spleen, and gastrointestinal tract; respiratory tract amyloidosis, however, is extremely rare. Due to a relative lack of typical clinical manifestations at this location, it is likely to be missed or misdiagnosed in the clinic.

Case Report

The patient described in this report provided written informed consent for publication. A 51-year-old woman (Figure 1) was admitted to hospital primarily due to intermittent hoarseness over the past 7 years that had become aggravated in the past month. One year previously, the hoarseness had become aggravated and was accompanied by foreign body sensation; however, the patient still did not devote much attention to the condition and underwent regular treatment. In the past month, the hoarseness had become aggravated and persistent, and was accompanied by sore throat, and breathing and swallowing difficulties; oral medication was ineffective. At this point, the patient was referred to our hospital. Her vital signs were as follows: blood pressure 110/70 mmHg, heart rate 80 beats/min, respiratory rate 18 breaths/min, temperature 98.06°F, and oxygen saturation 92% on room air. The patient exhibited pharyngeal lymphoid follicular hyperplasia, and uvula located in the center, without bilateral tonsil swelling or hyperemia of the epiglottis. She exhibited a normal degree of lifting of the soft palate. Botryoidal masses were visible at the entrance of the throat, with clear lung sounds. There were no rhonchi or moist rales, heart size was normal and heart beat was regular. Her abdomen was soft, and there was no tenderness or rebound tenderness. The liver and spleen were impalpable, and edema and varicose veins were visible in both lower extremities.

Routine biochemical analysis results were as follows: osmotic pressure 307.1, aspartate amino transferase/alanine amino transferase ratio 1.3, high-density lipoprotein 1.65 mmol/L, apolipoprotein A1 11.776 g/L, and apolipoprotein B 0.519 g/L. Sputum culture, sputum smear, serum carcinoembryonic antigen, anti-nuclear antibody, extractable nuclear antigen, lambda light chain, kappa light chain, hepatitis B and C viruses, HIV and Treponema pallidum tests were all normal.

Chest computed tomography (Figures 2a and 2b) revealed several prominent features, including eccentric soft tissue density shadows in the tracheal bifurcation of the proximal segment of the trachea. Electronic laryngoscopy revealed grape-like masses in the right lower torus tubarius (Figures 3a and 3b). Botryoidal masses were detected in the throat. Electronic bronchoscopy revealed extensive new biological invasive growth from the nasopharynx to the glottis (Figures 4a-4k). With regard to bone marrow aspiration, hypoplasia was decreased mildly.

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Discussion

Amyloidosis encompasses a heterogeneous group of diseases characterized by the deposition of congo philic amyloid fibrils in the extracellular matrices of tissues and organs. There are two major types of amyloidosis: primary and secondary. Primary amyloidosis most commonly affects the respiratory system [3] Respiratory amyloidosis has been described in three forms: diffuse interstitial, nodular parenchymal, and tracheobronchial [4]. Amyloidosis occurring in the respiratory system may involve the larynx, trachea, bronchus, lung, and mediastinal lymph nodes. Respiratory tract amyloidosis may occur in cases of systemic amyloidosis, but more often occurs in isolation [5]. Primary laryngo-tracheobronchial amyloidosis is a rare pulmonary disease; laryngeal amyloidosis accounts for benign laryngeal tumors in approximately 0.2% to 1.2% of cases [6]. Tracheobronchial amyloidosis represents 0.5% of all symptomatic tracheobronchial lesions [7,8]. The main symptoms of laryngeal amyloidosis are hoarseness and difficulty breathing [9]. It may be accompanied by bleeding and severe respiratory obstruction. Because respiratory tract amyloidosis occurs in different parts of the respiratory tract, its clinical manifestations are different. In the upper airway, it is associated with symptoms of hoarseness, dyspnea, wheezing, and cough. In the lower airway, associated symptoms include hemoptysis, post obstructive atelectasis, and recurrent pneumonia.

The gold standard for diagnosis of primary laryngo-tracheobronchial amyloidosis is pathological analysis using Congo red staining that produces green birefringence under cross-polarized light [10]. In addition to macroscopic findings on bronchoscopy, involvement of the membranous trachea and calcification of the airways on computed tomography are useful findings that suggest the presence of disease [11]. Additionally, unequivocal typing of amyloid deposits is a key step in the management of these diseases. Presently, immunofluorescence and immunohistochemistry are widely used. Light chain amyloidosis is the most common type of systemic amyloidosis, and appears to be more common than previously believed [12,13]. To date, 36 distinct proteins have been identified as amyloid fibril proteins in humans [14]. Major organ involvement, disease prognoses, and treatments can differ among the different subtypes of amyloidosis.

Conclusion

There is currently no uniform standard treatment for primary laryngo-tracheobronchial amyloidosis, and therapies are limited, including surgery, laser surgery, endobronchial silicone stent implantation, radiation, chemotherapy, and oral dexamethasone [15]. Bronchoscopic recanalization therapy options include laser resection, bronchoscopic dilation and stenting, and cryosurgery. This may be useful for the proximal or mid-airway [16]. Because plasma cells are radiosensitive, low-dose radiation therapy remains the mainstay of treatment for tracheobronchial amyloidosis [17]. High-dose chemotherapy combined with autologous stem cell transplantation confers better prognosis in systemic light chain amyloidosis lesions [18]. Immunosuppressive agents and radiotherapy are ineffective in laryngeal amyloidosis [19]. Surgery, especially CO₂ laser-assisted laryngeal microsurgery, is the most important treatment for this disease, which aims to retain normal organization [20]. Respiratory amyloidosis is a rare disease; nevertheless, it is important to improve our understanding of the disease to develop more effective treatments.

References


