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Case report

Chyloptysis causing plastic bronchitis

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A B S T R A C T

Chyloptysis is a rare clinical problem that is associated with conditions affecting lymphatic channels in the thorax. Diagnosis is usually made when the patients present with expectoration of milky-white sputum or of thick tenacious mucus in the shape of smaller bronchi (bronchial cast). Typically the symptoms resolve after coughing up of the bronchial casts. Pleural, mediastinal, pulmonary or lymphatic abnormalities result in chyloptysis. Lymphangiography and detection of lipids (cholesterol or triglycerides) in sputum help to establish the diagnosis. However, lymphangiography may not be positive in all patients. We report 2 patients with chyloptysis and bronchial casts with different etiologies. Abnormal lymphatics were demonstrated in one of our cases, but the second patients lymphangiogram was normal. In this patient we suspect that high venous filling pressures due to congestive heart failure had a causative effect in the setting of compromised lymphatic drainage in the thorax due to a prior history of radiation therapy to the chest for lymphoma.

Introduction

Chyloptysis is an uncommon medical condition where chyle accumulates in the sputum [1]. The sputum is of a milky-white consistency, which may help in early diagnosis. Chyloptysis may lead to the formation of bronchial casts. Frequently, chyloptysis is caused by abnormal lymphatic drainage that results in discharge of chyle into the tracheobronchial tree [2]. Chylothorax, resulting from blockage of the lymphatic system due to radiation, trauma or malignancy, can also lead to the development of chyloptysis when a bronchopleural fistula is present [2]. Lymphoma is a common cause of chylothorax with Non-Hodgkin's lymphoma accounting for the majority of cases that are non-traumatic [3]. Other etiologies of chyloptysis include lymphangioma [4], lymphangiectasis [5], lymphangiomatosis [6], lymphangiomyomatois [7], yellow nail syndrome [2], and Behcet's disease [8]. Here we describe 2 cases of chyloptysis; first due to duplication of thoracic duct and the other, we believe, due to decompensated congestive heart failure in the setting of compromised lymphatic drainage as a result of mantle field radiation.

Case 1

A 32-year-old Caucasian female presented with a three-month history of severe episodic wheezing, cough and extreme dyspnea associated with a feeling of drowning. She had no benefit from bronchodilators and her spirometry was normal. Each episode was associated with expectoration of "white tree branches" (Fig. 1, Panel A) and subsequent resolution of her symptoms without any treatment. Bronchoscopy showed white milky secretions emanating from the right bronchus intermedius. Bronchial lavage was markedly positive for 'Oil-red O stain'. Lymphangiography revealed duplication of thoracic duct with a smaller duct on the left side (Panel B and C, black arrows), a prominent thoracic duct on the right side (Panel B: white arrow) and collection of lymphatic vessels below the carina (Panel C: white arrow). Thoracic duct ligation and removal of abnormal subcarinal lymphatics resulted in resolution of her symptoms for eight years of follow up.

Case 2

A 37-year old woman was seen with chief complaints of wheezing, shortness of breath and "cloudy" sputum production. She reported bringing up thick branching sputum when she was in decompensated heart failure (Fig. 2). She was diagnosed with non-Hodgkin's disease when she was 22 years old that was treated with ABVD chemotherapy and mantle field radiation. Her radiation treatment was complicated by premature coronary artery disease, prosthetic mitral valve placement, restrictive cardiomyopathy by transthoracic echocardiography and right heart catheterization. She was also found to have a heart block requiring pacemaker placement. For her coronary artery disease she required multiple

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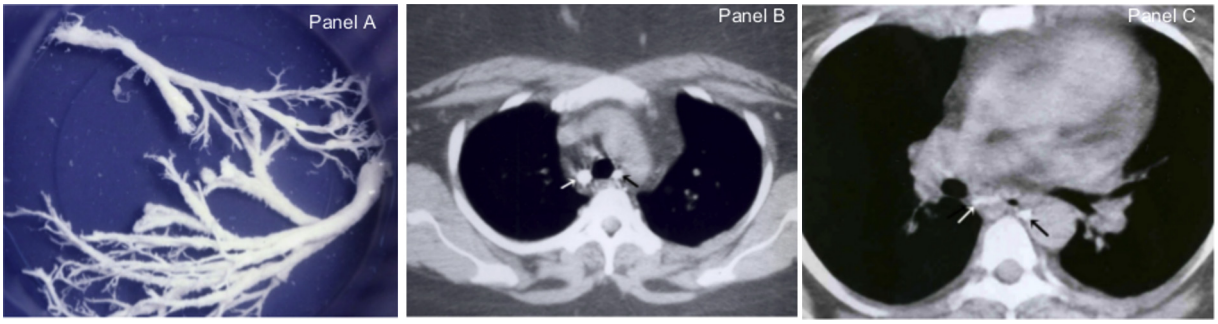

Fig. 1. Findings supporting the diagnosis of chylopysis- Panel A shows the 'white tree branches' like bronchial casts expectorated by patient in case 1. Lymphangiography revealed duplication of thoracic duct with a smaller duct on the left side (Panel B and C, black arrows), a prominent thoracic duct on the right side (Panel B: white arrow) and collection of lymphatics below the carina (Panel C: white arrow).

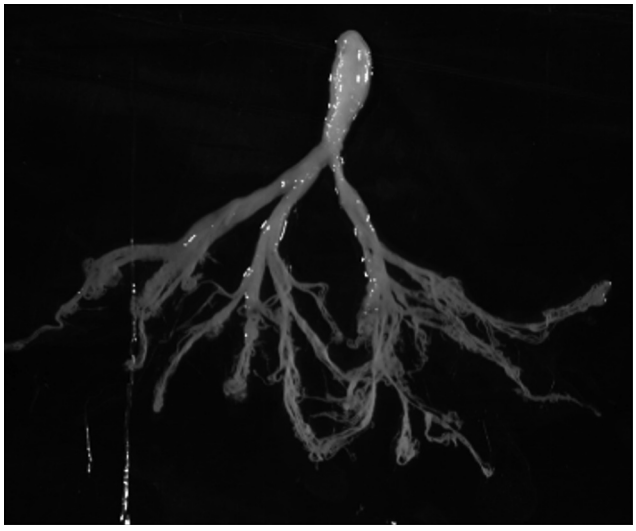

Fig. 2. A bronchial cast expectorated during periods of decompensated heart failure.

percutaneous coronary interventions and subsequently a coronary artery bypass grafting. Evaluation for bronchial casts included lymphangiography to evaluate for abnormal lymphatics and support the clinical diagnosis of chyloptysis, but it was non-diagnostic. Attempts to do lipoprotein electrophoresis failed due to the thickness of the sputum. She was aggressively treated for her cardiomyopathy and it was noted that she did not experience bronchial casts as long as she was optimally treated for heart failure. She coughed up small pieces of bronchial casts when she was volume overloaded with pulmonary edema on imaging studies. She was treated with mucolytics and nebulized bronchodilators with some response. In this case, a diagnosis of chyloptysis and bronchial casts was made based on clinical history and an extremely convincing history of plastic bronchitis. We believe that the etiology of chyloptysis and bronchial casts were related to decompensated CHF with underlying compromised lymphatic drainage due to history of non- Hodgkin's lymphoma and mantle field radiation.

Discussion

Chyloptysis is an uncommon clinical entity which has been associated with a number of conditions. We present two cases of chyloptysis with different etiologies. Case 1 presented with duplication of the thoracic duct and an abnormal connection to the airway. Case 2 had a history of Non-Hodgkin's lymphoma, treated with chemotherapy and mantle field radiation, and congestive heart failure with high filling pressures as well as worsening symptoms of plastic bronchitis associated with progressive heart failure.

Chyloptysis primarily arises from disruption of lymphatic drainage in the thorax. Chlye from the lymphatic system enters the bonchii either by a direct connection between the airway and the lymphatics or from the pleural space in presence of the bronchopleural fistula. Lymphatic channels from the lungs enter the right and left bronchomediastinal trunks. These drain into several locations including the subclavian and internal jugular veins on their respective sides, the right lymphatic duct, and the left thoracic duct. The thoracic duct carries chyle from the gastrointestinal system that is rich in triglycerides, chylomicrons and cholesterol [9]. Thoracic duct obstruction via trauma, or malignancy [2,9] can lead to an accumulation of chyle in the pleural space causing a chylothorax and chyloptysis. A common cause of malignancy related chylothorax is non-Hodgkin's lymphomas [3]. In presence of a chylothorax, a bronchopleural fistula is required for to chyle move into the tracheobronchial tree. Chylous mucus in the airway can solidify overnight resulting in formation of chylous bronchial casts [2]. Even in absence of anatomical obstruction, underlying cardiac abnormalities are a potential contributory factor for bronchial cast formation as in our second case. High venous pressure from cardiac defects or untreated heart failure can potentially cause impaired lymphatic flow into the venous system. In the presence of abnormal lymphatic connections to the airway, the lymph will likely flow into the airways leading to the development of bronchial casts.

Case reports of chyloptysis in the literature describe abnormal lymphatics including an irregular thoracic duct with multiple lymphatic channels branching off the thoracic duct [2] as well as thoracic lymphangiectasis [5]. Malignancy is associated with lymphatic obstruction either by way of direct obstruction by the tumor or as a result of the radiation treatment. A case of lymphangioma presenting with a chylous pericardial effusion is one example [4]. Additional case reports highlight disorders associated with chyloptysis including yellow nail syndrome [4] and Bechet's disease [8] Also, coronary artery disease [2] is documented in the literature as a condition presenting with chyloptysis.

Chyloptysis is primarily diagnosed by lymphangiography or CT lymphangiography [2]. Tests to detect fat content, such as Oil-Red O staining, can be carried out on the bronchial casts or bronchial alveolar lavage fluid. Bronchoscopy can be performed to look for an irregular connection between the pulmonary and lymphatic system. Patients presenting with lymphangioma [4] lymphangiectasis

[5], lymphangiomatosis [6], lymphangiomyomatois [7] yellow nail syndrome [2], and Behcet's disease [8] should be evaluated for chyloptysis if presenting with chylous sputum. Although our second case had a non-diagnostic lymphangiography, we believe that the lymphatic drainage was compromised due to mantle field radiation, and she experienced chyloptysis and bronchial casts when she was in decompensated heart failure which further compromised the drainage of lymphatics due to high filling venous pressures. Alternatively, collaterals may have developed that contributed to the symptoms.

Medical treatment for chyloptysis is focused on treating the primary cause and decreasing chyle formation. Measures to decrease chyle production include a low in fat diet supplemented with medium chain triglycerides which results in bypassing the thoracic duct [10] thus decreasing the amount of chyle draining into the bronchi. Management of the bronchial casts includes bronchoscopic extraction, therapy to aid with the expectoration of the casts, and medication including mucolytic drugs, bronchodilators, antibiotics, and steroids [11]. Surgical treatment for chyloptysis includes ligation of the thoracic duct as well as surgical removal of obstructed lymphatic vessels or pleurodesis if chylothorax and bronchopleural fistula are contributing to the development of chyloptysis.

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