Case Report

Plastic bronchitis in beta thalassemia minor

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ABSTRACT

Plastic bronchitis is a rare pulmonary disorder associated with various conditions like cystic fibrosis, asthma, pulmonary infection and characterized by formation and expectoration of cast which assumes the shape of the bronchial tree. We report a case of a 33-year-old woman with beta thalassemia minor who developed plastic bronchitis.

KEY WORDS: Beta thalassemia minor, bronchial casts, plastic bronchitis

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INTRODUCTION

Plastic bronchitis (PB) has also been called "fibrinous bronchitis," "pseudomembranous bronchitis," "Hoffman's bronchitis," and "cast bronchitis." It has been diagnosed in a variety of pulmonary pathologies including asthma, respiratory infections, cystic fibrosis, bronchiectasis, and acute chest syndrome associated with sickle cell disease, thalassemia alpha.[1,2] Several non-pulmonary pathologies[3] have also been associated with the diagnosis of plastic bronchitis. These include pericardial effusion, heart failure, post-Fontan surgery. Plastic bronchitis has not been reported in association with thalassemia minor. Although the disease is uncommon, its importance is based on the fact that early diagnosis has improved prognosis.

CASE REPORT

A 33-year-old woman with beta thalassemia minor was hospitalized with productive cough, dyspnoea with wheezing, and chest pain predominantly in interscapular area, ongoing since six months. There were no symptoms of asthma since childhood, or atopy in self or family. The patient was treated with empirical anti-tuberculous treatment without any relief, and then was referred to our side. The patient was previously diagnosed case of beta thalassemia minor as a part of workup done for her daughter who has Thalassemia major. General examination was normal. Respiratory system examination showed movements reduced on left with signs of volume loss, dull note on percussion, and reduced intensity of breath sounds over left hemi thorax. Blood hemogram was within normal limit. Sputum for acid fast bacilli smear and culture was negative. Chest radiograph [Figure 1] showed left lung collapse which was confirmed on computed tomography [Figure 2]. Bronchoscopy showed stenosed left mainstem bronchus just distal to carina with viscid secretions. The secretions were successfully aspirated. Her bronchial washings and post-bronchoscopy sputum for cytology were negative for malignant cells. Histopathology of the biopsied stenosed site was suggestive of respiratory epithelium with mucous. The patient was treated with oral corticosteroids, bronchodilators, *N*-acetyl cysteine nebulization, and vigorous chest physiotherapy for 3 months. In the mean time the patient continued to expectorate bronchial casts [Figure 3]. Post-treatment there was evidence of response noted clinically by improvement in intensity of breath sounds, radiologically by re-expansion of base of left lung confirmed on computed tomography [Figure 4] which showed aeration of upper and lower lobe with persistent post-obstructive collapse of the lingula.

DISCUSSION

PB is a rare disease characterized by the formation of large gelatinous or rigid airway cast. These casts are large and more cohesive than those seen in ordinary mucus plugging. Seear *et al.*,[4] proposed that bronchial casts can be divided into two distinct groups: Type 1: Inflammatory casts are

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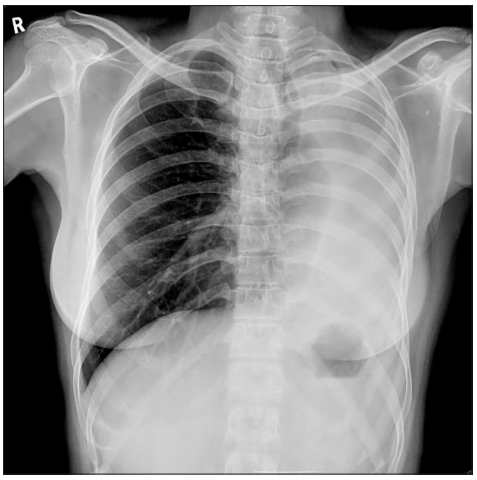

Figure 1: Chest radiograph showing collapse of the left lung

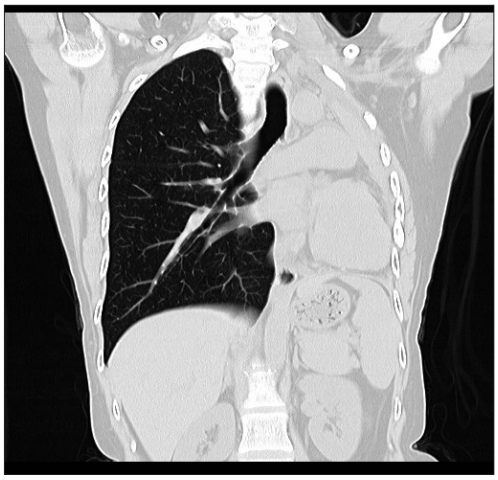

Figure 2: Axial reconstruction of CT Thorax showing complete collapse of the left lung

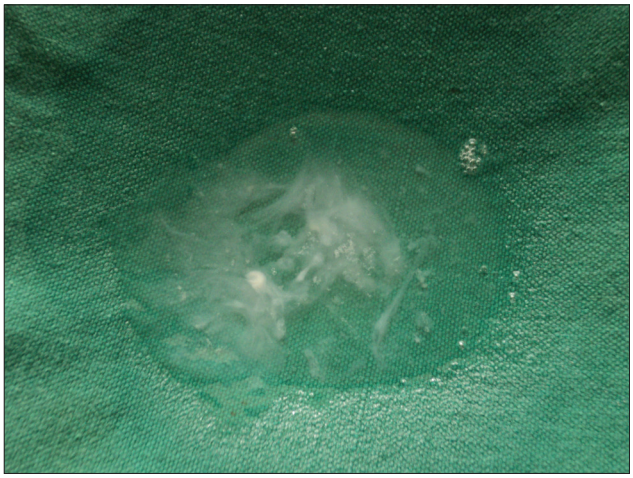

Figure 3: Coughed out viscid fibrinous material (casts)

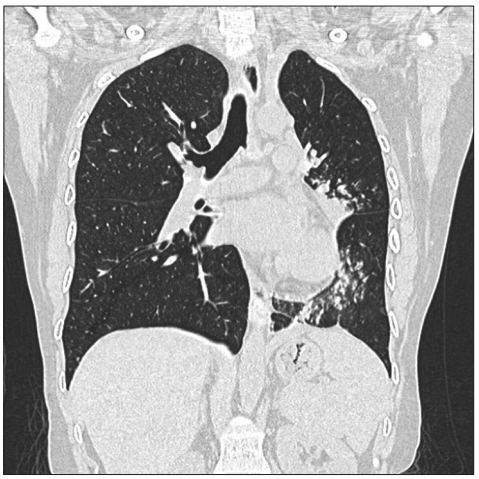

Figure 4: Post treatment CT Thorax showing aeration of upper and lower lobe with persistent post obstructive collapse of the lingula

composed primarily of fibrin with little mucin and have cellular infiltrates, particularly comprised of eosinophils. Type 2: Acellular casts are composed mainly of mucin with little fibrin and no inflammatory cells except occasional mononuclear cells. This was histologically similar to our case. Some patients with PB may have pathological features of both groups. A revised classification has been proposed depending on clinical disease association and cast histology.[5]

The pathogenesis of PB is not well understood. There are likely two mechanisms for the development of plastic bronchitis[5] (i) injury to bronchi and/or impaired bronchial epithelial function secondary to inflammation or infection e.g., in asthma, bronchiectasis, cystic fibrosis, sickle-cell anemia and (ii) impaired pulmonary lymphatic drainage. Association of beta thalassemia minor has been coincident. Since PB has been reported with hematologic disorders like sickle cell anemia and thalassemia alpha due to infections, infarcts, and sequestration, anemia which is a known phenomenon in beta thalassemia minor can lead to the above physiological abnormality.[2,6,7] They are known to present variedly, from being asymptomatic to having an acute chest syndrome.[2] Infection and inflammation increase the content of fibrin and cellular components in the casts. The specific feature of PB is the formation of obstructive bronchial plugs or casts of thick, tenacious mucus during an attack of bronchitis, when one or more lobes or even an entire lung may collapse. Casts are often expectorated,[3] but they may be discovered only at bronchoscopy, or be found lying in the bronchial tree at necropsy.

The treatment of PB is focused on the mechanical removal of the thick bronchial casts and prevention of further cast formation, removal of bronchial casts and maintenance of adequate ventilation, chest physiotherapy, bronchodilators, *N*-acetyl cysteine,[8] repeated bronchoscopic aspiration. Steroids have been used widely to treat PB with certain underlying pathologies. Marked clinical and radiological improvement in a patient with PB is secondary to its anti-inflammatory action.[9]

Similar clinical presentation and radiological pictures should alert a physician to the likelihood of malignancy

as plastic bronchitis is an uncommon disorder, but it can mimic malignancy-related lung collapse. If a physician suspects PB and subjects the patient to an immediate flexible bronchoscopy, etiologic diagnosis can be made earlier and the prognosis of patients with this condition can be improved. Although PB has been reported in association with thalassemia alpha, it has not been reported in association with thalassemia minor. Association of PB in beta thalassemia minor appears to be due to increased susceptibility to infection in these patients.[6,7]

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