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CASE REPORT

Bronchiolitis obliterans

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Abstract

The term 'bronchiolitis obliterans' was historically used by pathologists to refer to two distinct patterns of small-airway disease. The first was characterized by intraluminal polyps in the small airways. It was subsequently named bronchiolitis obliterans with organizing pneumonia and, more recently, cryptogenic organizing pneumonia. The second pattern was characterized by sub-epithelial inflammatory and fibrotic narrowing of the bronchioles, which is now recognized as obliterative bronchiolitis or constrictive bronchiolitis. In this study we reported a case of a male patient of 55 years who got admitted to a teaching hospital with progressive dyspnea and nonproductive cough over a period of 12 months and was released after proper treatment and management.

Key words: Bronchiolitis obliterans, fluticasone propionate, montelukast, treatment outcome.

Introduction

Bronchiolitis obliterans is lung disease characterized by fixed airway obstruction, inflammation and scarring occurs in the airways of the lungs resulting in severe shortness of breath and dry cough.[1,2] Bronchiolitis is a disease of the small airways, which are defined as airways less than 2 mm in diameter, and without cartilage.[3] The bronchioles are especially vulnerable to infectious or inhalational insults because of their small diameter. The primary symptoms of bronchiolitis are cough, dyspnea wheezing and feeling tired. Pulmonary function test reveals a nonreversible obstructive ventilatory defect and hyperinflation and often reveals diffusion impairment. Results of chest radiography may be normal. High-resolution computed tomography (CT) of the chest is the imaging study of choice, and characteristic findings include areas of gas trapping, bronchial-wall thickening, and centrilobular nodules.[4] Bronchiolitis may occur as a distinct clinical entity, such as acute bronchiolitis or obliterative bronchiolitis (also known as constrictive bronchiolitis), or as part of an interstitial lung disease with bronchiolar involvement, such as respiratory bronchiolitis-associated interstitial lung disease.[5,6]

It is important to identify this uncommon case of progressive dyspnea and dry cough to provide early and appropriate management and to provide appropriate explanation to the patient, and with that aim we reported a case of a male patient of 55 years who got admitted to a teaching hospital with progressive dyspnea and nonproductive cough over a period of 12 months, abnormal pulmonary function characterized by an obstructive airflow pattern and air trapping.

Case report

A 55 year-old man hailing from Khulna was

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Table 1. Laboratory findings

Investigation	Measurement
Haemoglobin	13.0 gm/dl
White blood cell	6.0 x 10^3/mm
Platelet	170.0 x 10^3/mm
Serum calcium	2.3 mmol/L
Serum albumin	38.0 gm/L
α_1-antitrypsin	1.2 gm/L
Rheumatoid factor	Negative
Serum creatinine	1.1 mg/dl
Serum glutamic pyruvic transaminase	22.0 U/L
Human immunodeficiency virus	Negative
Antibodies to cyclic citrullinated peptide	Negative
Anti-neutrophil cytoplasmic antibodies	Negative
Bronchoalveolar lavage fluid from the right middle lobe and lingula	Appearance: Clear Neutrophil: 36.0% Lymphocyte: 22.0% Monocyte: 2.0% Eosinophil: 8.0% Macrophages: 32.0% *Cytology*: Negative for virus, fungi or malignant cells. *Gram staining*: Very few mixed gram-positive and gram-negative organisms, and cultures were negative for bacterial, viral, and fungal pathogens.
Pulmonary function test (forced expiratory volume in one second, FEV_1)	*Before bronchodilator*: FEV_1 55% *After bronchodilator*: 62% of predicted
Chest X-ray	Normal
High-resolution CT scan	Mild cylindrical bronchiectasis predominantly in the lower lobes, diffuse bronchial-wall thickening, no difference in the attenuation of the lung parenchyma on expiratory images compared to inspiratory images, which suggested air trapping.
Lung biopsy and histopathology	*At low magnification*: Absence of bronchioles in most of the specimens. *At higher magnification*: Respiratory bronchioles were encircled by collars of dense fibrosis and fibrous and elastotic scarring, with virtual obliteration of the lumen. Linear scars lay along the bronchioles, along the cross section, the findings were consistent with a diagnosis of obliterative bronchiolitis with fibrosis.

admitted to a teaching hospital with the complaints of progressive shortness of breath which was initially with exertion and subsequently interferes with his activities of daily living and non productive cough persisting for last 12 months.

Over last 12 months, the patient suffered from febrile illness for 4 times. All episodes were associated with shortness of breath and dry cough. Every time the patient was treated at a local hospital with intravenous antibiotics, steroid inhaler and oral montelukast. The patient was discharged with long-term steroid inhaler and oral montelukast. Despite regular medication, the patient felt it difficult to complete activities of daily living without being short of breath. For that reason he got admitted to a teaching hospital for better management.

He had no known allergies to medications. He was married and had two adult children. He was a worker in a battery manufacturing company. He lived in a suburban area, drunk safe water and used sanitary toilet. His father had chronic obstructive pulmonary disease (COPD), brother had dilated cardio myopathy, and children were healthy.

On examination, the temperature was 99°F, the blood pressure 125/73 mmHg, the pulse 78 beats per minute, the respiratory rate 16 breaths per minute, no clubbing and the oxygen saturation 93% while he was breathing ambient air. The body-mass index was 19 kg/m^2. The lungs were clear, and the remainder of the examination was normal.

Results of pulmonary-function tests are shown in Table 1. The hemoglobin, white blood cell count, platelet count, and blood levels of calcium, albumin, and α1-antitrypsin were normal, as were results of testing for rheumatoid factor and tests of renal (serum creatinine) and liver function (serum glutamic pyruvic transaminase). Testing was negative for human immunodeficiency virus (HIV), antibodies to cyclic citrullinated peptide, and anti-neutrophil cytoplasmic antibodies.

Bronchoalveolar lavage fluid was drawn from the right middle lobe and lingula. The appearance was clear with neutrophil count 36.0%, lymphocyte 22.0%, monocyte 2.0%, eosinophil 8.0% and macrophages 32.0%. The cytology was negative for virus, fungi or malignant cells. The gram staining showed very few mixed gram-positive and gram-negative organisms, and cultures were negative for bacterial, viral, and fungal pathogens.

The pulmonary function test revealed forced expiratory volume in one second (FEV_1) 55.0% before bronchodilator and 62.0% of the predicted after bronchodilator.

The finding of chest X-ray was normal. The high-resolution CT scan showed mild cylindrical bronchiectasis predominantly in the lower lobes and diffuse bronchial-wall thickening. There was no difference in the attenuation of the lung parenchyma on expiratory images compared to inspiratory images, which suggested air trapping.

Lung biopsy and histopathological examination revealed an apparent absence of bronchioles in most of the specimens at low magnification, and showed respiratory bronchioles encircled by collars of dense fibrosis and fibrous and elastotic scarring, with virtual obliteration of the lumen at higher magnification. Through the cross section, bronchioles showed the linear scars consistent with a diagnosis of obliterative bronchiolitis with fibrosis.

Discussion

Cause and diagnosis

Etiological factors includes collagen vascular disease, transplant rejection in organ transplant patients, viral infection (respiratory syncytial virus, adenovirus, HIV, cytomegalovirus, exposure to toxic fumes including sulfur dioxide, nitrogen dioxide, ammonia, chlorine, thionyl chloride, hydrogen chloride, mustard gas.[7,8] In some cases it can be idiopathic.[9-11] Some industrial workers are more at risk namely nylon flock worker, spray print worker in textile, battery workers and workers who use or manufacture flavorings.[12] Symptoms may not occur until 2 to 8 weeks following toxic exposure or infec-

tions. The patient of this study was a worker in a battery manufacturing company and suffering from this disease for long time.

Diagnosis of bronchiolitis obliterans can be challenging due to its mimicking of asthma, chronic bronchitis, emphysema and pneumonia.[7-11]

Numbers of diagnostic tools are used to reach the correct diagnosis includes chest X-ray, diffusing capacity of the lung test, spirometry (fixed airway obstruction), lung volume test (hyperinflation), high resolution CT (air trapping, thickening of airway), and lung biopsy.[9-11]

Treatment

The patient was counseled that the disease is irreversible and severe cases often require lung transplantation, and transplant recipients are at risk for re-developing the disease as bronchiolitis obliterans is a common complication of chronic rejecter.[9] A study has shown the combination of fluticasone propionate, oral montelukast, and oral azithromycin may stabilize the disease and slow the disease progression.[12] The patient was treated with intravenous levofloxacin, oral azithromycin, nebulized fluticasone propionate and oral montelukast. The patient became stable and suggested ambulatory treatment and follow-up visit when condition deteriorates.

Prognosis

The prognosis is often poor, most people die in a month to years. Long term definitive treatment is lung transplantation.

Prevention

Prevention can be done by improving exhaust system in workplace, proper using of personal protective equipment, monitoring potentially affected workers and improving awareness on the disease among industrial workers through health education.[13]

Conclusion

Bronchiolitis obliterans is not very uncommon but can easily be missed and misdiagnosed. A case has been reported in this study. This might be helpful to clinicians to be careful that bronchiolitis obliterans as a differential in a patient with long term shortness of breath and dry cough which are poorly responsive to the management of bronchial asthma, COPD or bronchiectasis.

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