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Case Report

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Adult Diagnosis of Swyer-James-Macleod Syndrome: A Rare Case Presentation

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Abstract

Swyer-James-MacLeod syndrome also known as unilateral hyperlucent lung syndrome is a rare entity associated with post infectious bronchiolitis obliterans occurring in childhood. It is characterized by hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion. The diagnosis is usually made in childhood but sometimes occurs in adulthood. The disease often presents with dyspnea, decreased exercise tolerance, cough, hemoptysis, and recurrent pulmonary infections. SJMS may be confused with asthma or pulmonary embolism due to similar symptoms and may result in inappropriate therapy.Here we report a case of 26 year old female presented with cough, exertional breathlessness and fever for the past 8 months.She was diagnosed as SJMS on the basis of her medical history, clinical presentation and x-rays and computed tomography chest scan findings.

Keywords: Bronchiolitis obliterans, Hyperlucentlung, Bronchiectasis, Atelectasis, Pneumonectomy

Introduction

Unilateral pulmonary emphysema as a clinicoradiological entity was described by Swyer and James in 1953[1] and by MacLeod in 1954[2].It is a relatively uncommon entity, occurring in 3.8% of patients with bronchiolitis obliterans (BO) in one study and in 4.3% of BO cases in another[3].Swyer-James-Macleod syndrome(SJMS), or unilateral hyperlucent lung syndrome, is a rare entity associated with post-infectious bronchiolitis obliterans occurring in childhood[4]. This syndrome is a long-term complication of bronchiolitis in children, especially after adenoviral infection occurring in infancy[5].

The affected child may be asymptomatic, but more often, the patient has recurrent pulmonary infections and develops bronchiectasis. A basic pathologic condition is bronchiolitis associated with obliteration of the small airways and a severe emphysematous pattern owing to related alveolar destruction and dilated lung parenchyma[6]. Peripheral pulmonary vascularization is decreased as a result of inflammation. Unilateral or bilateral involvement is possible. There are various proposed mechanisms for the presence of bronchiectasis in SJMS including BO leading to atelectasis or scarring which results in

bronchial dilatation, bronchiectasis itself being the primary inciting event with distal spread to peripheral small airways leading to obliteration, or the initial viral infection may damage the bronchioles and bronchi simultaneously[3].

This disorder is typically diagnosed in childhood after an evaluation for recurrent respiratory infections, but patients who have little or no sequelae of bronchiectasis sometimes have minor symptoms or are asymptomatic and may therefore not be diagnosed until adulthood[7].

Case Presentation

A 26 year old female patient admitted in emergency with complaints of breathlessness, cough with scanty sputum production and fever for the past 3 months. Her symptoms had started after a severe infection of the respiratory tract at the age of 9 years. The patient's symptoms decreased and disappeared from time to time but continued for 17 years. She had been treated for bronchial asthma with bronchodialators and steroids previously. She was also started on antituberculous treatment from private hospital on clinicoradiological basis. She had no history of smoking. On examination patient was dyspnoeic, with a blood pressure 100/70, pule rate 108/minute, respiratory rate 30/minute and saturation was 84% on room air. On general physical examination pallor and clubbing were present. Respiratory examination revealed decreased breath sound on left side with bilateral coarse crepitation and bilateral ronchi.

Laboratory findings showed a white cell count of 13800, Haemoglobin 7.7g/dl ESR 5mm/hr RBS 152mg/dl, Serum Bilirubin 1.5,SGOT 350,SGPT 795,S ALP 221. Her serum electrolytes and renal function test were within normal ranges. Her sputum examination for acid fast bacilli and fungus were negative. The chest radiograph disclosed a hyperlucent left lung with numerous cystic regions of bronchiectasis (Fig. 1). Chest CT scans demonstrated pulmonary artery hypoplasia, widespread bronchiectasis with a small calibre left pulmonary artery and volume loss in the left lung. (Fig. 2). Echocardiography revealed severe pulmonary artery hypertension with dialated right atrium (Fig3). Abdominal ultrasound was normal.

Fig.1

Fig.2

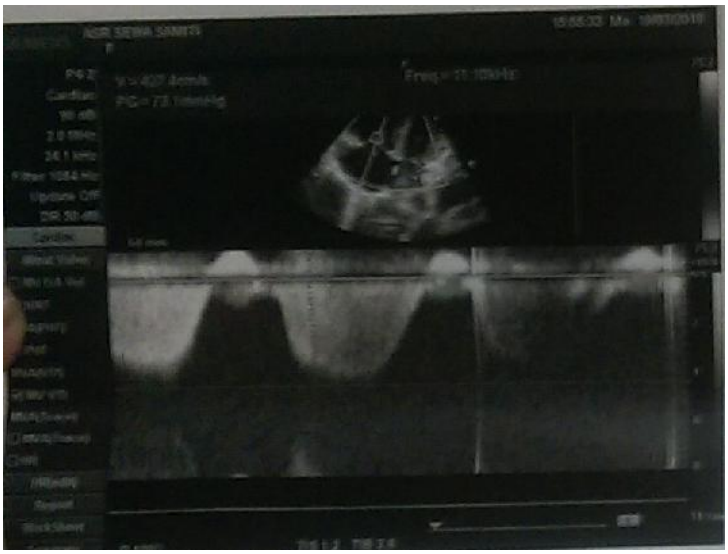

Fig.3

Discussion

SJMS also known as unilateral hyperlucent lung syndrome which was first described in 1953 and 1954 is a rare entity characterized by hyperlucency of one lung, lobe or part of a lobe. There has been much debate regarding the exact etiology of this process however it is associated with childhood infections most commonly post-infectious bronchiolitis obliterans and pneumonitis. It is also associated with various viruses such as Paramyxovirus morbillivirus, *Bordetella pertussis*, Mycobacterium tuberculosis, *Mycoplasma* pneumoniae, influenza A and adenovirus types 3, 7 and 21[8] .This patient gives history of pneumonia in childhood.

Unilateral bronchiolitis obliterans with hyperinflation is quite rare, with a prevalence of 0.01% in 17,450 surveyed chest radiographs[9]. Clinically, the disease often presents with dyspnea, decreased exercise tolerance, cough, hemoptysis, and recurrent pulmonary infections[5]. Dyspnea on exertion was the most frequent symptom in a series with 8 patients in contrast to an earlier study of 9 patients in which dyspnea was not a prominent feature[10].

In our case patient had most of these symptoms. Dyspnea and cough were the most frequent symptoms. Clubbing may be due to the presence of bronchiectasis.

Abba and Al-Mobeireek found that most adults with SJMS are symptomatic, often for a prolonged period of time at presentation[3]. This patient had ongoing symptoms for nearly 17 years at intervals.

Complications of unilateral hyperlucent lung syndrome include recurrent infection in areas of bronchiectasis, lung abscesses, and spontaneous pneumothorax[11,12].

Diagnosis is made radiographically by x-ray and CT scan and is an incidental finding in some cases. Radiographically, the hyperlucency is usually confined to one lobe or lung. In this case chest x ray showed left side hyperlucency with normal sized lung, decreased broncho-vascular markings and a small hilar shadow. Thorax CT and high-resolution CT provide useful additional information, such as patchy bilateral regions of hyperlucency or bronchiectasis. In this case CT showed the typical findings of hyperlucent areas with diffuse oligemia and air trapping. Bronchiectasis is not necessary for diagnosis but is present in some cases, and there are several explanations for this. First, bronchiolitis obliterans can cause atelectasis or scarring, which in turn leads to bronchial dilatation.

Treatment is usually individualized ranging from conservative management to surgical intervention. Conservative symptomatic management is the mainstay of treatment for patients diagnosed with SJMS utilizing chest physiotherapy, low-dose inhaled corticosteroids, and inhaled bronchodilators[13]. Patients should also be given pneumococcal and Influenza vaccinations. Long-term oxygen therapy may be appropriate in cases with advanced disease and respiratory failure[14]. Surgical intervention should be considered for patients who have repeated infections and are not responding to optimal medical management. The most common surgical procedure was a pneumectomy and some patients were treated with lobectomy or segmentectomy[13]. Prognosis is dependent on the presence or absence of bronchiectasis[14].

There are several important differential diagnoses that should be considered when evaluating any individual with unilateral pulmonary hyperlucency including pneumothorax, asymmetric emphysema, congenital lobar emphysema, and pulmonary artery hypoplasia[14].

Other differential diagnoses include gastrointestinal herniation, bronchial compression, mastectomy and mediastinal fibrosis[15]. Poland syndrome is another cause of a unilateral hyperlucent hemithorax, which is due to congenital unilateral absence of the pectoralis major and minor muscles, hypoplasia of the breast and nipple and scarcity of subcutaneous tissue[16]. SJMS can be easily misdiagnosed and must be suspected in any patient diagnosed with asthma who does not respond to therapy. One case highlighted this, as their patient was initially misdiagnosed as having asthma[17].

Conclusion

The main reason for reporting this case series is the rarity of patients who have been diagnosed with SJMS in adulthood. Also it is important to remember the differentials of unilateral lung hyperlucency as inappropriate diagnosis can lead to inappropriate therapy.

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