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Case Report

SWYER JAMES SYNDROME- A COMPLICATION OF CHILDHOOD LUNG INJURY

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ABSTRACT
Swyer James Syndrome also known as Macleod syndrome is hyperlucency of one lung, lobe or part of lobe due to pulmonary vascular abnormalities and alveolar over distension. Primary etiology is childhood respiratory infection which leads to obliterative bronchiolitis. We hereby report this rare syndrome in a 20 year old Asian female who presented with complaints of breathlessness on exertion and cough with expectoration. She had history of childhood pneumonia. On examination she had diminished breath sounds and hyper resonant percussion note on left side of chest. Chest radiology and computed tomography showed emphysema and diminished vascular markings on left side and mediastinal shifting towards right side. Based on history, clinical examination and radiology patient was diagnosed as Swyer-James Syndrome.

Keywords: *Swyer James Syndrome, Macleod Syndrome, Obliterative Bronchiolitis, Emphysema*

CASES
20 year old female, belonging to low socioeconomic status from South Delhi, India, presented with breathlessness on exertion for 2 years and cough with expectoration for 15 days.
Detailed history revealed that her breathlessness was insidious in onset and progressed from MMRC dyspnea grade 1 to grade 2 at present. There was no orthopnea, no paroxysmal nocturnal dyspnea (PND). She had cough which was associated with white colour, non fowl smelling, small amount of sputum. There was no history of haemoptysis. She didn't have fever, chest pain. Her appetite was fair and there was no significant weight loss.
Past & Personal History:
She had recurrent history of rhinitis due to deviated nasal septum at least twice in a year which use to subside with course of antibiotics and intra nasal anti allergic drugs.
There was history of childhood pneumonia (patient is not able to recall detail childhood history). No history of tuberculosis in past. No history suggestive of diabetes, hypertension. No significant relevant history in family.
On Examination:
General examination: no significant abnormal findings.
Systemic Examination: ENT- Deviated Nasal Septum towards right side. No other significant abnormality.
Respiratory system: Both side normal vesicular breath sound present but breath sound decreased in intensity on left side significantly. Hyperresonant percussion note on left side.
Other system examinations were clinically normal.

Investigations

1. Routine blood: TLC- 18,000, DLC-N-80%, L-15%, E-3%. RBS- 126 mg%, ESR- 56 1st hour. B.Urea- 20mg%, S.Creatinine-0.9mg%
2. Blood culture- no bacterial growth.
3. Sputum Culture- pseudomonas aeruginosa.
4. Sputum Acid fast bacilli- Negative
5. Serum alpha-1 antitrypsin levels were within normal limits.

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6. Chest X ray- Left side emphysema with mediastinam deviated towards right side.
7. Computed Tomography Thorax (CT Thorax): Pan lobular and bullous emphysema of left lower lobe with marked air trapping, relative paucity of vascular markings on left side, mediastinal shift to right, partial loss of volume of right lung.
8. Spirometry- FVC- 65%, FEV_1 – 80% FEV_1/FVC ratio- 0.9. suggestive of Restrictive pattern.
9. Bronchoscopy: all airways are patent, no intraluminal growth.

Final Diagnosis: Swyer James syndrome with secondry lower respiratory tract infection with deviated nasal septum.

Treatment and Clinical course: Patient was given 2 weeks of antibiotic course according to drug sensitivity report along with inhaled bronchodilators. On followup Patient was asymptomatic with treatment but still had breathlessness on exertion. Later on patient was referred to ENT surgeon for deviated nasal septum correction.

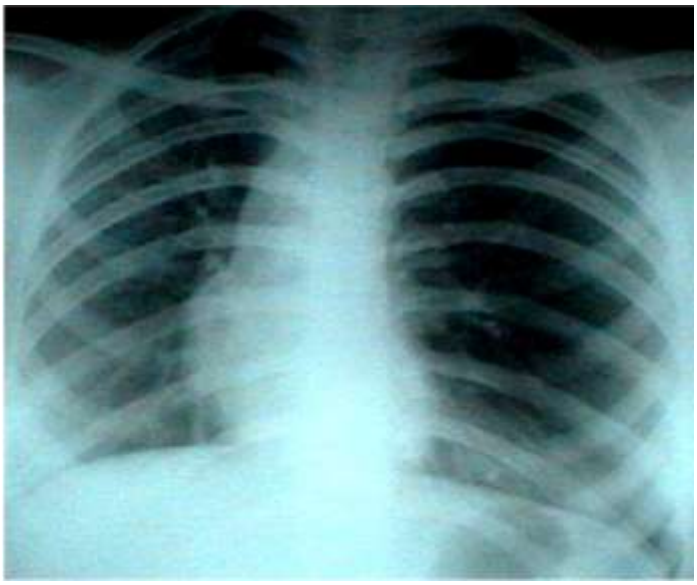

Figure 1: Chest X ray shows Hyperlucency of left lung. Mediastinal shifting towards right side

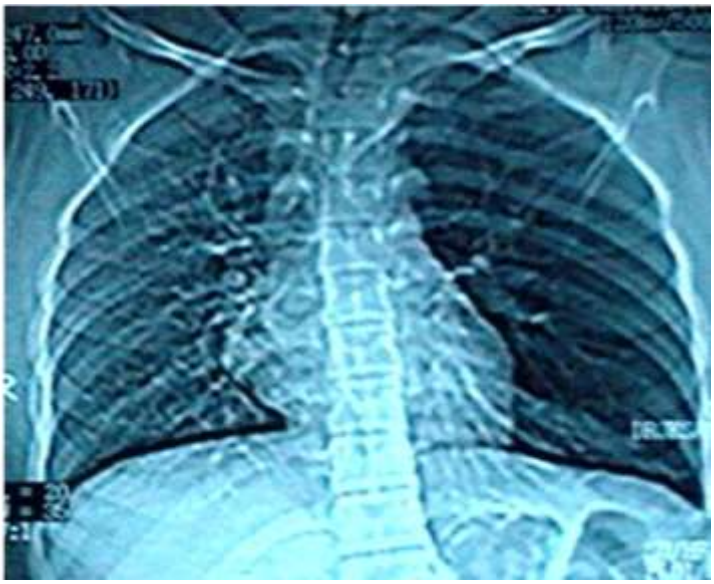

Figure 2: CT THORAX shows left side emphysema with Paucity of vasculature on left side

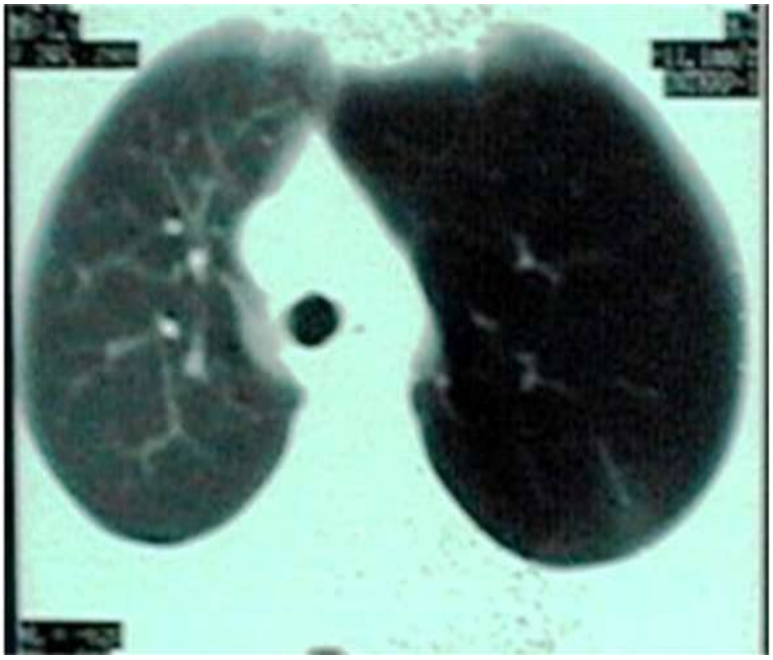

Figure 3: CT Scan thorax shows pan lobular and bullous emphysema of left side, relative paucity of vascular markings on left side, mediastinal shift to right and partial loss of volume of right lung

RESULTS AND DISCUSSION

Swyer-James syndrome also known as MacLeod syndrome is an uncommon disorder which was first described in 1953. Among 52 patients with bronchiolitis obliterans, Gosink *et al.*, (1973) reported only two cases of hyperinflation. Fregonese *et al.*, (2002) reported prevalence of 0.01% in a survey of 17,450 chest radiographs.

SJS is characterized by unilateral hyperlucency of lung with unilateral reduction in vascularity (Quaedvlieg *et al.*, 2004; Chan *et al.*, 1999). Due to unknown factors, it usually involves the left lung (Abba and Al-Mobeireek, 2003). Etiology is unknown but it is a form of obliterative bronchiolitis with concomitant vasculitis following injury to immature lungs during the first 8 years of life. Common cause of lung injury are due to childhood respiratory infections like measles, whooping cough, Tuberculosis, Mycoplasma pneumonia, influenza A, adenoviral infections etc (Schlesinger *et al.*, 1998) and other non-infective causes like aspiration, toxic fumes, and organ transplantation (McLoud *et al.*, 1986). Both small bronchi and bronchioles are affected and the lung with abnormal airways remains inflated by collateral air drift (Collins and Stern, 1999). This damage during the early childhood prevents normal development of the alveolar ducts. Airways develop submucosal fibrosis leading to luminal irregularity and occlusion. Pulmonary vasculature is hypoplastic while the lung distal to diseased bronchioles become hyperinflated and sometimes pan acinar emphysematous changes develop (Collins and Stern, 1999).

Patients with little or no bronchiectasis have minor symptoms like sputum production and breathlessness, or are asymptomatic and may remain undiagnosed until adulthood. Moreover, adult patients with SJS are often diagnosed after a chest radiograph obtained for another reason (Smeds and Fortune, 2011).

Pulmonary function test shows restrictive pattern most of time but obstructive pattern may be present due to associated bronchiectasis (Collins and Stern, 1999).

The diagnosis of SJS requires the exclusion of other causes of unilateral hypertranslucency like pneumothorax, asymmetric emphysema, congenital lobar emphysema (CLE), pulmonary artery hypoplasia, pulmonary embolism and bronchial obstruction due to foreign body or mucus plugs (Tortajada *et al.*, 2004). Hyperlucency in this case is due to reduced perfusion and air trapping. Pulmonary vessels are reduced on the affected side but lung volumes are only slightly decreased or same. Ipsilateral air trapping is a key finding (Collins and Stern, 1999).

The characteristic radiographic findings in these patients include unilateral hyperlucent lung along with decreased broncho vascular markings (Gopinath *et al.*, 2005).

Despite characteristic findings by chest radiography, Computed Tomography (CT) of thorax is the imaging technique of choice in establishing diagnosis of SJS (Lucaya *et al.*, 1998). CT scan findings of Swyer-James syndrome include patchy areas of low attenuation and hypovascular areas interspersed with areas of normal attenuation. Air trapping is confirmed on expiratory scans. Other changes on CT may include bronchiectasis, atelectasis and scarring (Moore *et al.*, 1992; Marti-Bonmati *et al.*, 1989).

Pulmonary ventilation-perfusion scintigraphy shows marked decrease in ventilation, vascular flow and perfusion in the affected regions (Mera *et al.*, 2006).

Treatment of SJS are primarily conservative and supportive. Antibiotic therapy, use of bronchodilators and chest physiotherapy with postural drainage of the secretions may be helpful.

Long-term oxygen therapy is required in cases with advanced disease and respiratory failure. Surgery in the form of lobectomy or pneumonectomy can be offered to those who had severe symptoms despite optimal therapy. Prognosis largely depends on presence of associated bronchiectasis (Quaedvlieg *et al.*, 2004; Gopinath *et al.*, 2005).

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