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Case Report

Adult diagnosis of hyperlucent unilateral lung: Swyer-James Macleod syndrome

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ABSTRACT

Swyer- James- MacLeod Syndrome (SJMS) is a constrictive bronchiolitis with airflow obstruction, decreased number and diameter of ipsilateral peripheral pulmonary vessels and air trapping with characteristic radiological feature of unilateral hyperlucency on chest radiography. Computerized tomography (CT) provides useful additional information. The diagnosis is usually made in childhood, but sometimes it in adult. Here, we report a case in a 36 year old adult female who presented with complaints of breathlessness on exertion, cough and recurrent rhinitis with past history of TB and child hood chest infection. Imaging findings are characteristic of Swyer- James- Macleod syndrome.

Keywords: Constrictive bronchiolitis, Swyer-James-MacLeod syndrome, Hyperlucency, CT

INTRODUCTION

In 1953, Swyer and James, in 1954 Macleod, giving much more detail, described patients with unilateral hyperlucent lung. Swyer-James-Macleod syndrome (SJMS) or unilateral hyper lucent lung syndrome is a rare entity associated with post infectious bronchiolitis obliterans occurring in childhood.[1]

This syndrome is a long-term complication of bronchiolitis in children, especially after adenoviral infection occurring in infancy.[2] The affected child may be asymptomatic, but more often patient has recurrent pulmonary infections and later develops bronchiectasis. Dyspnoea, haemoptysis, and chronic productive cough are visible symptoms. A basic pathologic condition is the bronchiolitis associated with the obliteration of the small airways and severe emphysematous pattern. Owing to related alveolar destruction and dilated lung Parenchyma.[3] The chest radiograph demonstrates lobar or unilateral hyper lucent lung; normal or reduced volume of the affected lung. Airflow obstruction is generally present on pulmonary function testing (PFT), and ventilation and perfusion (V/Q) scanning often reveals markedly decreased perfusion of the affected lung.[4] The bronchographic findings are striking and limited to the abnormal lung.[5] The major bronchi are normal, but the smaller branches are club-like, and occasionally small buds project from the ends of the peripheral divisions. Generally, there is almost complete absence of alveolar filling, demonstrated by a well-demarcated clear zone between the bronchiectatic smaller bronchi and the chest wall.[5] This disorder is diagnosed in childhood after an evaluation for recurrent respiratory infections typically, but sometimes patients who have little or no bronchiectasis have minor symptoms or are asymptomatic and may, therefore, miss their diagnosis until adulthood.[6] The rarity of the disorder and imaging curiosity prompts us to report this case.

CASE REPORT

A 38 year old female presented to department of respiratory medicine, with complains of shortness of

breath on exertion, which progressive in nature. She had cough with mild expectoration and is non-foul smelling. There was no history of haemoptysis. She did not have fever, chest pain. She had recurrent history of rhinitis due to allergy which subsides with course of antibiotics and intra nasal anti allergic drugs. She was diagnosed asthmatic in 2004 on treatment but symptoms are not relived completely. She had tuberculosis in 2002 for which she had taken treatment. There was history of childhood infection. No history suggestive of diabetes, hypertension. General examination reveals no significant abnormal findings. Respiratory examination revealed reduced chest movements and breath sounds on the left hemithorax with crackles in the left lower lung fields and bilateral rhonchi. Other system examinations were normal. She had normal haematological and biochemical parameters. Pulmonary function tests revealed moderate airway obstruction.

Radiological Investigations

Chest radiograph shows small left lung with slight hyperlucency on left side as compared to right lung. Mediastinal shifted towards left side. There are also few calcified nodular opacities seen in bilateral lung field suggestive old sequelae of tuberculosis.

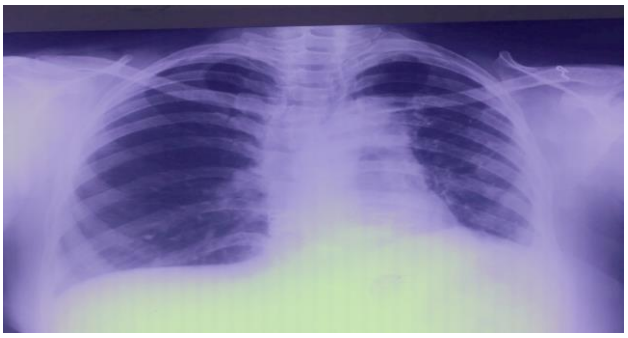

Figure 1: Small left lung with slight hyperlucency on left side as compared to right lung. Mediastinal shifted towards left side. There are also few calcified nodular opacities seen in bilateral lung field suggestive old sequelae of tuberculosis.

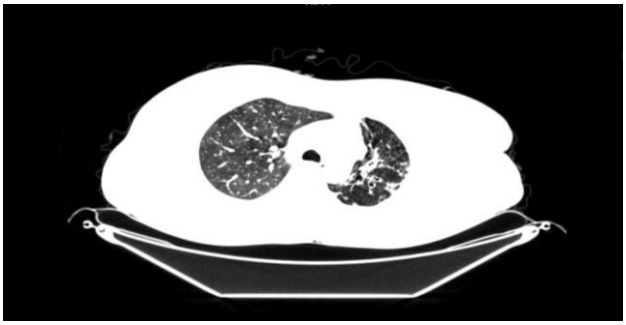

Figure 2: CT axial image in lung window – left upper lobe shows bronchiectatic changes with fibrotic lesion and small areas of air trapping in form of hypertranslucency.

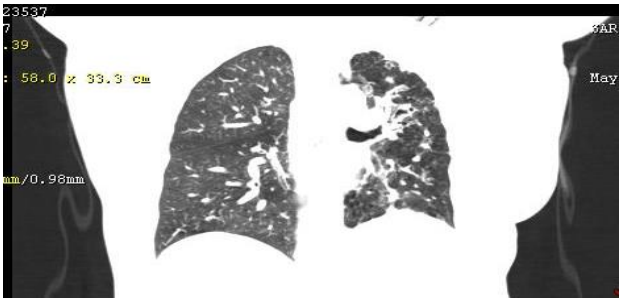

Figure 3: Computed tomographic coronal reformat image showing reduce volume of left lung with areas of increased translucency

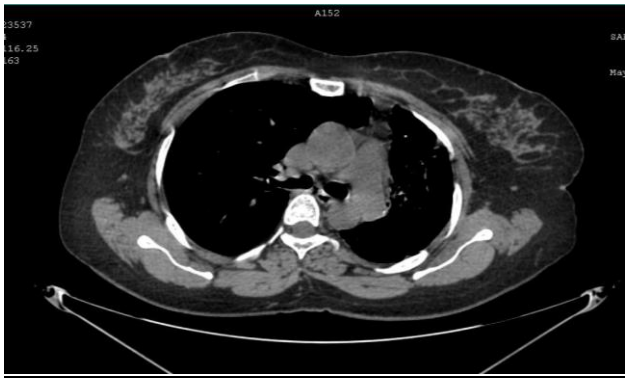

Figure 4: Computed tomographic axial image in mediastinal window shows small calcific foci in left upper lobe with fibrotic changes - sequelae of tuberculosis (Post ATT).

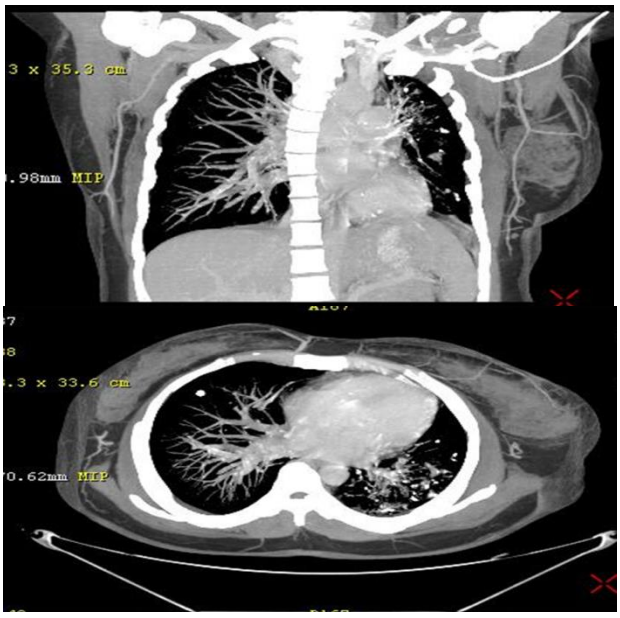

Figure 5: Computed tomographic coronal reformat (a) and axial (i) imaging in mediastinal window (MIP) - in comparsion to right lung, left lung shows slightly reduced calibe of left pulmonary artery with paucity of left pulmonary artery branches. It also shows calcific foci.

Computed tomography of chest confirms that left lung volume is reduced with focal area of hyper translucency suggestive of air trapping. Multiple fibro nodular lesions are seen in left lung parenchyma with bronchiectatic changes in left upper lobe. Few small calcified nodules are also seen in right lung. Left lung shows decreased vascularity with reduced caliber and paucity of left pulmonary artery branches. Mediastinal is shifted towards right side.

DISCUSSION

Unilateral bronchiolitis obliterans with hyperinflation is quite rare, with prevalence of 0.01% in 17.450 surveyed chest radiographs.[7] It is presently considered to be an acquired disease secondary to viral bronchiolitis and pneumonitis in childhood etiological associated with *Paramyxovirus morbillivirus, Bordetella pertussis, Mycobacterium tuberculosis, Mycoplasma pneumoniae, influenza A* and *adenovirus* types 3, 7 and 21.[1]

Clinically, the disease is often presents with dyspnoea, decreased exercise tolerance, cough, haemoptysis and recurrent pulmonary infections.[2] In present case she had one or more of these symptoms. Physical examination findings are non-specific and may include decreased chest expansion, rales or hyper-resonance. Abba's study showed that most adults with SJMS are symptomatic, often for a prolonged period of time at presentation.[9] Complications of unilateral hyperlucent lung syndrome include recurrent infection in areas of bronchiectasis, lung abscess, and spontaneous pneumothorax.[10,11]

Swyer-James-Macleod syndrome (SJMS) is characterized by unilateral hyperlucency of lung with unilateral reduction in vascularity.[8] Due to unknown factors, it usually involves the left lung.[9] 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 and other noninfective causes like aspiration, toxic fumes, and organ transplantation.[12,13]

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 panacinar emphysematous changes develop.[14]

Patients with little or no bronchiectasis have minor symptoms like sputum production and breathlessness, or are asymptomatic and may remain undiagnosed until adulthood. Pulmonary function test shows restrictive pattern most of time but obstructive pattern may be present due to associated bronchiectasis.[14] The diagnosis of SJS requires the exclusion of other causes of unilateral hypertranslucency of lung are given below (Table 1).[15]

Pulmonary vessels are reduced on the affected side but lung volumes are only slightly decreased or same. Ipsilateral air trapping is a key finding.[14] These adult patients with SJMS are often diagnosed after a chest radiograph obtained for another reason. The classic chest radiographic finding is a pronounced one-sided hyperlucency due to oligaemia of the involved segments of the lung.[16]

A mediastinal shift toward the affected side may occur on inspiration. Expiratory radiograph may demonstrate air trapping or a shift of the mediastinum towards the unaffected side. Bronchiectatic changes may also be observed on chest radiographs. Despite characteristic findings by chest radiography, CT is the imaging technique of choice in establishing the diagnosis of SJMS.[17]

Table 1: Causes of unilateral hypertranslucency of the lung.

	Causes of unilateral hypertranslucency of the lung
Normal	Increased density of contralateral lung, e.g. pleural effusion/thickening, consolidation
Technical	Rotation, scoliosis
Soft tissue	Mastectomy; Congenital absence of pectoralis muscle Poliomyelitis
Emphysema	Obstructive: foreign body, tumour, Macleod's syndrome, congenital lobar; Emphysema bullous; Compensatory: lobar collapse, lobectomy
Vascular	Obstructed pulmonary artery, e.g. by tumour, embolus; Absent/hypoplastic; pulmonary artery; Macleod's syndrome
Pneumothorax	Others

On CT, SJMS appears as hyperlucent areas due to decreased pulmonary perfusion of the lung without an anteroposterior gradient attenuation. As stated earlier, the hyperlucency may not be confined to one lung. In addition, the distribution of hyperlucency within the involved lung may be inhomogeneous with patches of normal lung attenuation demonstrable. Ventilation/ perfusion scans are an important modality in the

diagnosis of SJMS. Ventilation/perfusion scans document matched ventilation and perfusion defect.[18]

CONCLUSION

As childhood respiratory infection in considered to be an important cause of SJMS. In present case, patient has no significant childhood respiratory infection history but patient had past history of tuberculosis and also diagnosed asthmatic on the basis of PFT. Same PFT findings may also coexist with SJMS. Other environmental and genetic factors should be considered in etiology. Therefore, relevant history with positive imaging findings leads to the correct diagnosis of SJMS.

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