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Swyer- James -MacLeod syndrome presenting as hemoptysis in an adult

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ABSTRACT

Swyer-James/MacLeod syndrome is an uncommon disease with characteristic radiological feature of unilateral hyperlucency due to loss of pulmonary vasculature and air trapping. Typically, this disorder is diagnosed in childhood during evaluations for recurrent respiratory infections. Here, we report a case in a 30-year-old adult female who presented with dyspnoea, cough with expectoration and recurrent hemoptysis due to associated bronchiectasis. This case highlights the importance of computed tomography in the diagnostic workup of recurrent hemoptysis in pulmonary tuberculosis epidemic countries like India.

Key-words:
Swyer-James/MacLeod syndrome, unilateral hyperlucent lung, hyperinflation, bronchiolitis obliterans, pulmonary artery abnormalities.

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Introduction:

Swyer-James/MacLeod syndrome was first described by Swyer and James[1] in 1953 and further detailed by Macleod in 1954.[2] It is a rare acquired disease occurring secondary to postinfectious bronchiolitis in childhood.[3] Bronchiolitis obliterans leads to reflex vasoconstriction and hypoperfusion causing hypoplasia of pulmonary vasculature leading the development of hyperlucent pathologic lung. Usually this disorder is diagnosed in childhood after an evaluation for recurrent respiratory tract infections but sometimes their diagnosis may be missed until adulthood if the patient has no or little sequelae bronchiectasis. The rarity of the disorder and clinical curiosity prompts us to report this case.

Case History:

A 30-year-old female presented to our outpatient department with complaints of dyspnoea, cough with expectoration for last twenty years and recurrent hemoptysis for the last three years. She was taking the treatment for asthma diagnosed on the basis of clinical and spirometry for last eighteen years. For the complaint of hemoptysis she was prescribed multiple courses of emperical antitubercular drugs in the past without sputum smear examination. She had normal haematological and biochemical parameters. On examination her vital signs were within normal limits. Respiratory examination revealed reduced chest movements and breath sounds on the left hemithorax with coarse crackles in the left lower lung fields and bilateral rhonchi. Once hemoptysis was controlled her sputum for acid fast bacilli was done twice which turned out to be negative. Her admission chest radiograph showed left sided hyperlucent lung with small ipsilateral hilar shadow [Figure 1]. A diagnostic bronchoscopy was done, but there was no endobronchial growth or bleeding points. As clinical and radiologic findings were unable to explain the cause of hemoptysis computed tomography (CT) of thorax was done which revealed small left lung with decreased vascularity and bronchiectactic segments in the left lower lobe with airtrapping on expiration [Figure 2a and 2b]. As CT was suggestive of the

disorder, a pulmonary angiogram was done to confirm the diagnosis which revealed hypoplasia of left pulmonary artery and vein [Figure 3]. Therefore the patient was diagnosed to have Swyer-James-MacLeod syndrome and the cause of hemoptysis was ascertained to be due to bronchiectasis of left lower lobe as confirmed by CT. Ventilation perfusion scan and surgical removal of bronchiectactic segments were advised to the patient, both of which were refused by the patient. She was prescribed conservative management for hemoptysis and is under our regular follow up.

Table 1.Causes of unilateral hypertranslucency of the lung [7].

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

Compensatory: lobar collapse, lobectomy
Obstructive: foreign body, tumour, Macleod's syndrome, congenital lobar emphysema
Bullous

Vascular

Absent/hypoplastic pulmonary artery
Obstructed pulmonary artery, e.g. by tumour, embolus
Macleod's syndrome

Pneumothorax

Discussion:

This disorder may present with various clinical features and the reported prevalence of this disease is 0.01% in 17,450 survey chest radiograph.[4] The patient can be completely asymptomatic with hyperlucent lung field as an incidental finding on the chest radiograph taken for other indications. Recurrent respiratory infections, productive cough, shortness of breath and dyspnoea on exertion and rarely hemoptysis as in our case may be the alternate presentations.[5] If there is no bronchiectatic component, the prognosis is usually favourable. The classic chest radiographic finding is a pronounced one sided hyperlucency due to the oligemia of the involved segments of the lung.[6] Expiratory radiograph may demonstrate air trapping or mediastinal shift towards the opposite side. The various causes of unilateral hyperlucent lung are given below [Table 1].[7] CT is the imaging technique of choice despite characteristic findings in chest radiograph.[5] It not only show hyperlucent areas, but also helps out in ruling out other causes of unilateral hyperlcent lung, endobronchial tumour partially obstructing the lumen of a lobe and in assessing the type and extend of bronchiectasis. This modality of investigation have a valuable role in countries like India were antitubercular drugs are first prescribed drugs for the management of hemoptysis without exactly knowing the etiology. Spirometry shows mixed pattern with reversible obstructive airway disease. Ventilation perfusion (V/Q) scanning of the lungs shows matched V/Q defects and a marked trapping on the washout phase on 133 Xe scintigraphy.[8] In our case despite doing various investigations the cause of hemoptysis in our patient was ascertained only until we did a CT.

In conclusion, this case reaffirms the utility of CT in diagnostic workup of recurrent hemoptysis in countries like India where tuberculosis is an epidemic.

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Figure 1.Admission chest radiograph (PA view) showing hyperlucent left lung(arrow) with small ipsilateral hilar shadow.

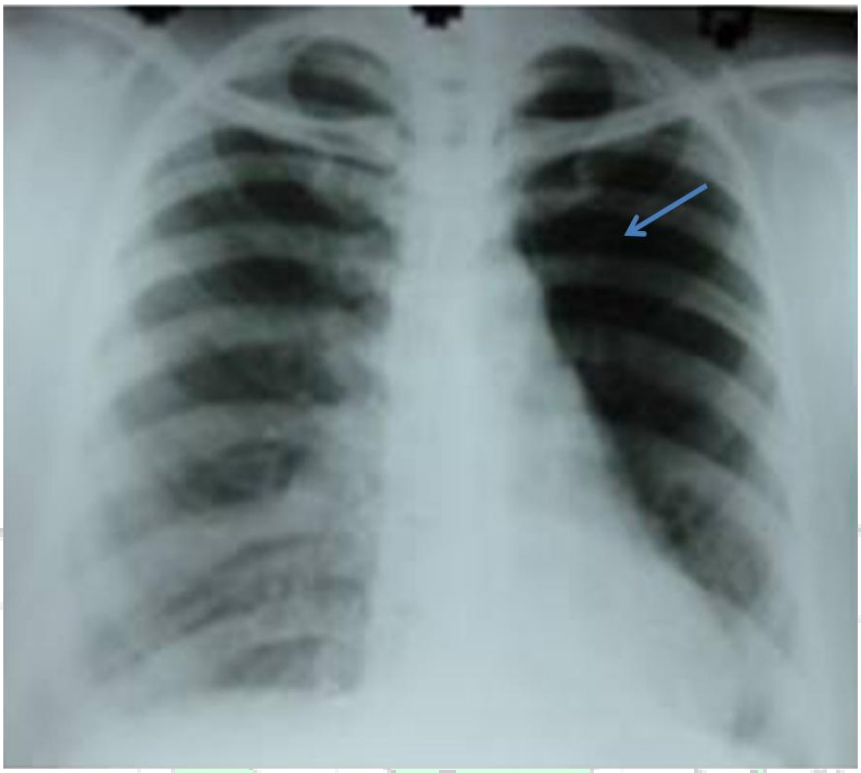


Figure 2:Computed tomography of chest (a) in mediastinal window showing small left pulmonary artery[blue arrow] and coronal CT image (b) showing hyperlucent areas in left lung field(Thick blue arrow) and bronchiectactic segments(Thin blue arrow) in left lower lobe.

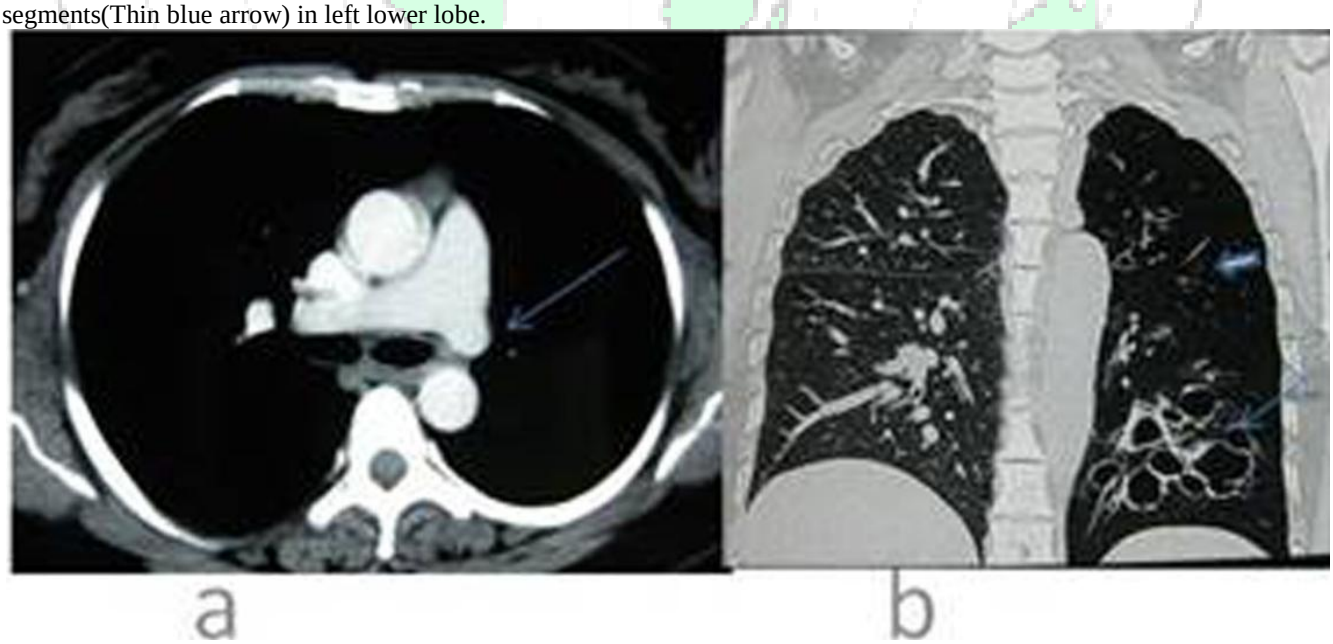
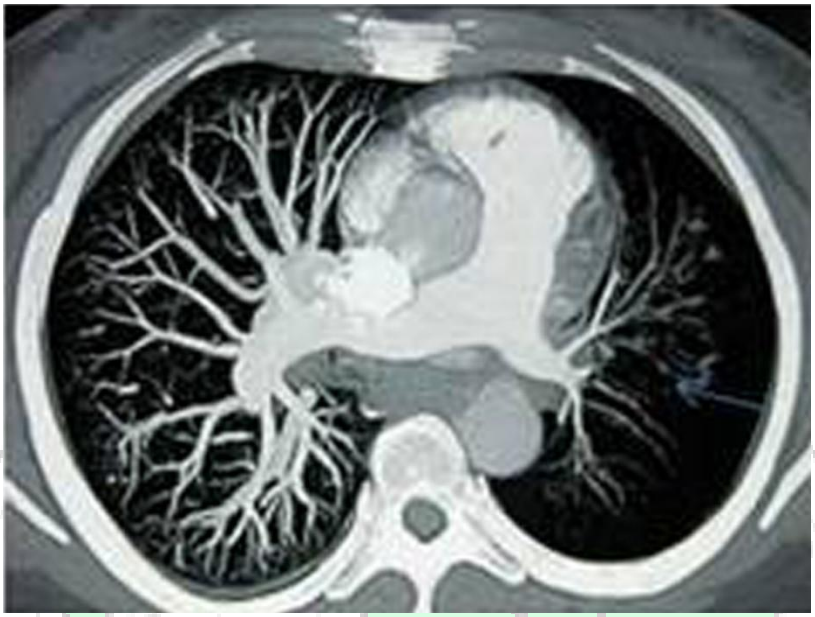


Figure 3: CT Pulmonary angiogram showing a small left pulmonary artery (arrow heads) with reduced size and number of branches with diminished peripheral vasculature in the left lung.



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