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A Rare Case of Swyer James Syndrome

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Abstract: ***Aims & Objectives:*** *To review the clinical presentation and imaging findings in a rare case of Swyer James Syndrome* ***Materials & Methods:*** *A 50 year old male patient presented with complaints of exertional dyspnea, cough with expectoration for the past three months. Detailed history revealed similar dyspnea episodes since childhood.* ***Result:*** *Patient underwent HRCT Thorax which revealed unilateral radiolucency in the left lower lobe with decreased vascularity with associated bronchiectasis and nodular opacities. Mosaic attenuation pattern with air trapping on expiratory scan was also seen. CONCLUSION: SJMS is considered to be a relatively uncommon and complex disease characterized by unilateral hyperlucency of a part of or the entire lung which was first described in 1953 by Swyer and James. Some patients, who have little or no associated sequelae bronchiectasis, have minor symptoms or are asymptomatic and may, therefore, not be diagnosed until they are adults. SJMS diagnosis is based on the radiological pattern such as unilateral or lobar pulmonary hyperlucency associated with an air trapping lung during expiration ultimately resembling a mosaic pattern. The affected lung parenchyma shows a variable degree of destruction and bronchiectasis could be associated. The diagnosis of this syndrome is better established with HRCT on inspiration and expiration complemented with an angio-CT*

Keywords: swyer james syndrome, macleods syndrome, air trapping, mosaic pattern, Bronchiolitis

1. Introduction

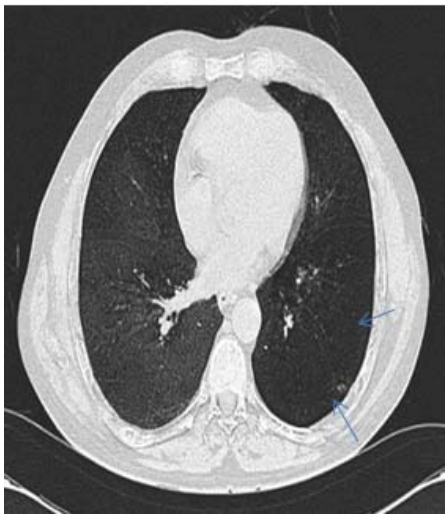
Swyer-James or *MacLeod syndrome* is a post-infectious constrictive bronchiolitis that is usually the sequelae of severe childhood pneumonia. Adenovirus types 3, 7, and 21 have been implicated most commonly, but other infectious etiologies include: *Mycoplasma pneumoniae*, parainfluenza virus types 1-3, influenza virus types A and B, respiratory syncytial virus, measles, and *Bordetella pertusis*. Unilateral transradiancy on plain chest radiography in Swyer–James syndrome reflects a combination of hypoplasia of the pulmonary vasculature and obliterative bronchiolitis. The affected lung is small or normal in volume.

2. Materials & Methods

A 50 year old male patient presented with complaints of exertional dyspnea, cough with expectoration for the past three months. Detailed history revealed similar dyspnea episodes since childhood. Patient underwent MDCT plain and contrast scan of thorax using 5.0 mm and 1.5 mm cuts. Inspiratory and expiratory scans were also taken using 1.0 mm cuts. volume rendering was also done to the images for better evaluation.

3. Results

Traction bronchiectatic changes in the inferior segment of left lingular lobe and antero-basal segment of right lower lobe and medial segment of right middle lobe. Subcentimetric centrilobular nodules in the subpleural region in the lateral basal and posterior basal segments of left lower lobe. Paraseptal emphesymatous changes in superior segment of right lower lobe. Mosaic attenuation with decrease in size of segmental pulmonary arteries (oligemia) in the left lower lobe. Air trapping seen on the expiratory scan in the left lower lobe.



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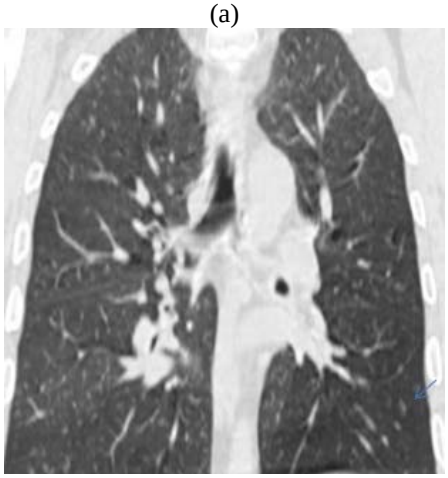

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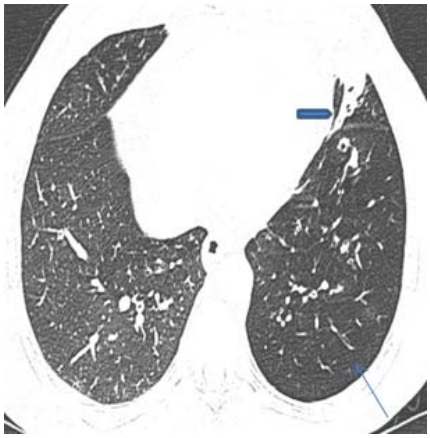

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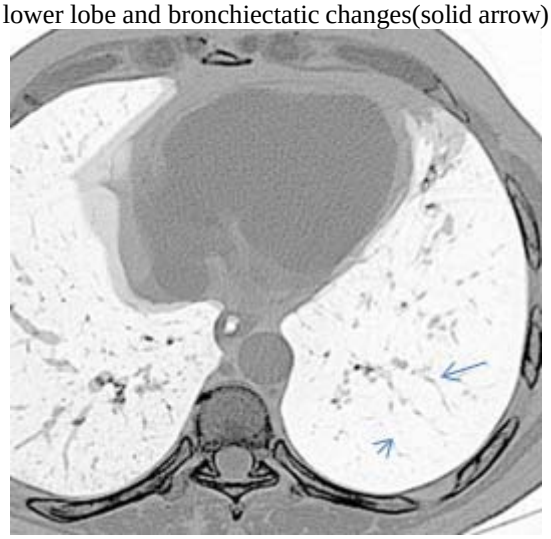
Figure 1: .a,b) axial and coronal hrct window shows mosaic attenuation at the level of left lower lobe(arrows) c) Expiratory scan axial views shows air trapping in the left lower lobe and bronchiectatic changes(solid arrow)



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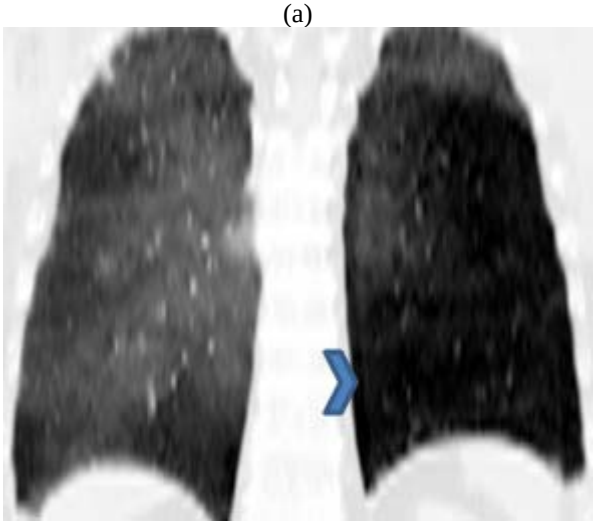

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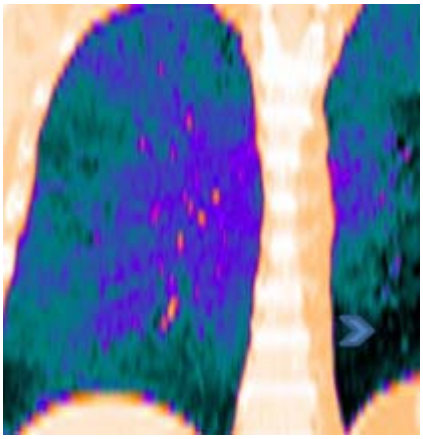

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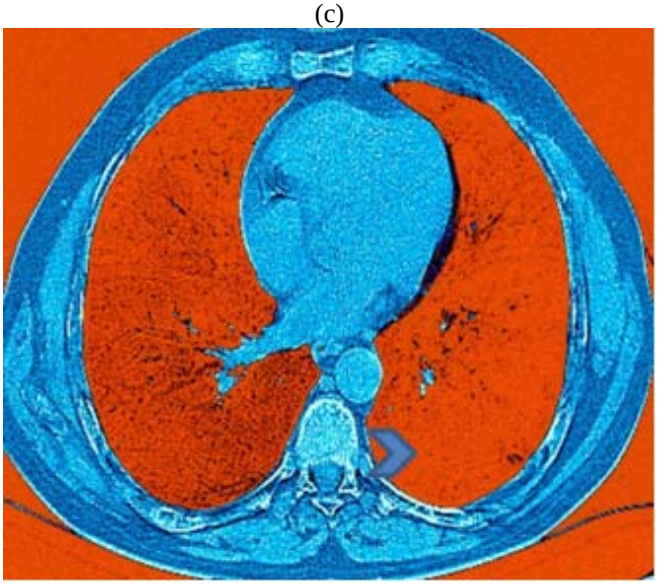

(d)

Figure 2: a) bone inversion axial image shows decreased segmental arteries at the level of left lower lobe (arrows) b,c,d)Mosaic attenuation seen on MIP and volume rendering images(arrow heads)

4. Discussion

The Swyer–James or McLeod syndrome is a particular form of obliterative bronchiolitis that has special features: it occurs following an insult to the developing lung. The lung served by damaged bronchi and bronchioles remains inflated by collateral air drift. As defined in the original descriptions, the disease on chest radiography is predominantly unilateral, giving rise to the key finding of unilateral transradiancy.

The condition is characterized by bronchitis, bronchiolitis, constrictive obliterative bronchiolitis, and probably emphysema.Typically the condition is unilateral and a whole lung is affected, but changes may be confined to a lobe or segment. Because it is usually an incidental finding on a chest radiograph performed for other reasons, there is no particular age at presentation. Other recognized patterns are of segmental sparing with the rest of the lung involved and of bilateral lobar or segmental disease. The patchy nature of lung involvement in some patients is particularly well demonstrated on CT examination.

Bronchi and bronchioles from the fourth generation to terminal bronchioles have submucosal fibrosis, causing luminal irregularity and occlusion. The lung parenchyma is hypoplastic, including the pulmonary artery and its branches, which are reduced in both size and number. Lung distal to diseased airways is hyperinflated and supplied by collateral air drift. Sometimes panacinar emphysematous changes are present, although the definition of emphysema in the context of developing lung is controversial.

The Swyer–James syndrome is caused by injury of the immature lung. Injury most commonly follows an acute viral infection occurring during the first 8 years of life, before the lung has completed its development. Viruses implicated include adenovirus and measles virus.

Nonviral causes include infections such as *Mycoplasma* pneumonia and pertussis. Patients are typically asymptomatic and commonly present as adults with an abnormal chest radiograph. Less commonly patients have exertional dyspnea, which may be progressive and, exceptionally, quite marked or repeated respiratory infections. When coincidental acute lung disorders occur in the presence of the Swyer–James syndrome, the chest radiograph may show a unilateral distribution of acute abnormality, as is recorded with pulmonary edema and pulmonary haemorrhage. Pulmonary function tests show a reduced vital capacity, airflow obstruction, and a reduced gas diffusing capacity.

Ipsilateral air trapping is a key finding and a *sine qua non* of the condition. It can be demonstrated on an expiratory radiograph which should be exposed during a forced expiratory manoeuvre because the short expiratory time maximizes volume differences between the obstructed and nonobstructed lung.

CT shows changes that are often more complex than suspected from the chest radiograph and while it may confirm that the radiographic transradiancy is largely one-sided, it more commonly shows bilateral abnormalities. Areas of decreased attenuation on CT are often inhomogeneous, containing a patchwork of local decreased attenuation and hypovascular areas interspersed with lung of normal density. Such small low-density areas may be poorly or sharply marginated, representing areas of small airways disease and air-trapping. Air-trapping can be confirmed with expiratory CT scans.

Other changes on CT include bronchiectasis, which is a frequent, but not universal, finding, and areas of collapse and scarring.
The described combination of radiographic findings usually allows exclusion of other conditions that may resemble the Swyer–James syndrome. These conditions include congenital hypoplastic lung, congenital lobar emphysema, pulmonary artery hypoplasia,and proximal interruption of the pulmonary artery. The greatest concern is that signs of Swyer–James syndrome are being produced by a central, large airway obstruction, causing lung hypoventilation and a compensatory ipsilateral reduction in perfusion. This is a problem that may be resolved only by bronchoscopy or a tailored CT examination of the central airways.

In contrast to the apparently unilateral distribution of Swyer–James syndrome on chest radiography, CT usually reveals bilateral abnormalities (areas of decreased attenuation and cylindrical bronchiectasis) in individuals with the syndrome.
Findings on the plain chest radiograph are characteristic. Unilateral transradiancy is caused by reduced lung perfusion. Lesser degrees of involvement are not easily detectable on the plain radiograph.

On the affected side the size and number of mid-lung and peripheral vessels are reduced. Blood flow in the contralateral lung is increased, and frequently this lung looks plethoric, an abnormality that may be more striking than the unilateral transradiance.

The hilum of the involved lung is small but lung volumes are normal or only slightly decreased. The mediastinum may show some shift to the affected side at total lung capacity. The fact that the ipsilateral lung volume is not increased is helpful in distinguishing Swyer–James syndrome from emphysema per se

5. Conclusion

SJMS is considered to be a relatively uncommon and complex disease characterized by unilateral hyperlucency of a part of or the entire lung which was first described in 1953 by Swyer and James. Some patients, who have little or no associated sequelae bronchiectasis, have minor symptoms or are asymptomatic and may, therefore, not be diagnosed until they are adults. SJMS diagnosis is based on the radiological pattern such as unilateral or lobar pulmonary hyperlucency associated with an air trapping lung during expiration ultimately resembling a mosaic pattern. The affected lung parenchyma shows a variable degree of destruction and bronchiectasis could be associated. The diagnosis of this syndrome is better established with HRCT on inspiration and expiration complemented with an angio-CT

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