

Macleod syndrome presenting as hemoptysis:

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Citation

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Abstract

Macleod syndrome is an important cause of unilateral lucency on chest radiograph and usually detected incidentally on imaging done for some other reason or sometimes with respiratory problems including infections or hemoptysis. We present a case of Macleod syndrome in an adult who presented with hemoptysis due to associated bronchiectasis.

INTRODUCTION

Macleod syndrome also known as Swyers and James syndrome is characterized on chest skiagram appearance of unilateral loss of lung volume with hyperlucency, unilateral reduction in vascularity on CT scan of the chest, and unilateral loss of perfusion on Technetium 99c lung scan and air trapping on expiration on imaging or ventilation scan using xenon [1234]. Its etiology is not well understood some authors believe in the congenital origin of the syndrome while some thinks it an acquired condition with broncho-pulmonary lesions being the primary abnormality and the pulmonary vascular hypoplasia developing later [1356].

Patients having this syndrome may be asymptomatic detected incidentally on imaging or may present with mild respiratory symptoms, recurrent pulmonary infections or hemoptysis [1278].

We report below a case of Macleod syndrome affecting the left lung of an adult who presented with hemoptysis.

CASE REPORT

A young female without any significant medical history presented with hemoptysis for the first time. On clinical examination no significant finding could be ascertained. She was initially treated on OPD basis but returned after 3 days with persistent hemoptysis. This was not associated with fever, loss of weight, loss of appetite or any other symptomatology. CXR done outside was reported as normal. Bronchoscopy revealed active bleed from left lower lobe, which was controlled by local administration of adrenaline.

On careful review of X-ray left lung appeared hypertransradiant with small ipsilateral hilar shadow

however cause of hemoptysis could not be explained based on X-ray chest (Figure- 1). Henceforth CECT and HRCT chest was done which revealed: -

- Small left lung with diminished vascularity
- Hypoplastic left pulmonary artery
- Bronchiectatic changes in posterior basal segment of left lobe (Figure- 2 & 3)

Figure 1

Figure 1: X-ray chest PA view shows hypertransradiant left lung with small left hilar shadow



Figure 2

Figure 2: CECT Chest in mediastinal window shows small left pulmonary artery

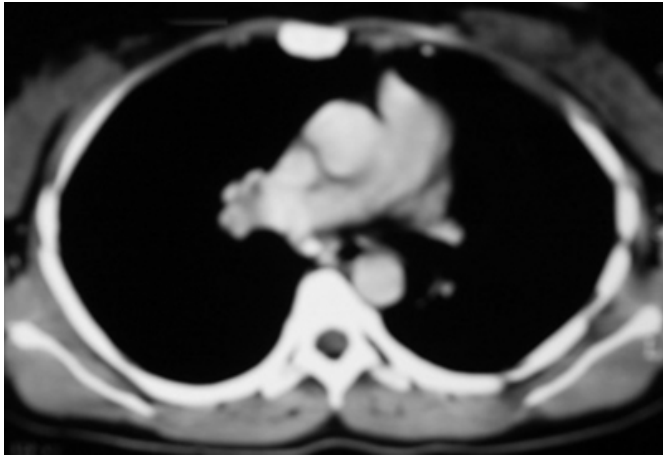
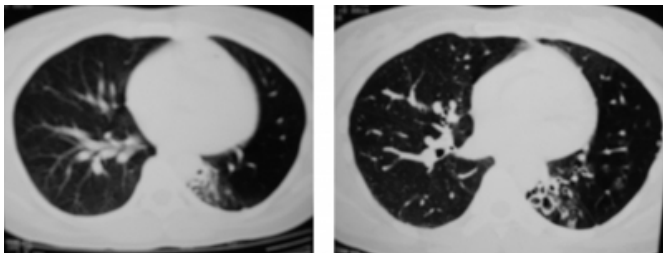


Figure 3

Figure 3: CECT and HRCT sections shows small left lung, diminished vascularity associated with cystic bronchiectasis in posterior basal segment of left lower lobe.



Sampling of the bronchial secretions showed *Nocardia* from the dilated bronchi – which probably precipitated the bleed.

Considering the radiological findings a possibility of Macleod syndrome with bronchiectatic changes was thought and diagnosis was confirmed when air trapping was demonstrated on expiratory skiagram (Not provided).

DISCUSSION

Macleod (1954) described nine adult cases of hypertransradiancy of one lung without collapse [9]. Swyers and James (1953) described a similar condition in a child [10]. Since then many cases both in children and adults have been reported. Some authors believe in the congenital origin of the syndrome with pulmonary artery hypoplasia being the primary defect [5]. Others however, believe that it is an acquired condition with broncho-pulmonary lesions being the primary abnormality and the pulmonary vascular hypoplasia developing later [610]. It is postulated that childhood viral infections causes bronchiolitis and obliteration of small airways; the involved distal airways are

ventilated by collateral air drift, and air trapping leads to panacinar emphysema [4]. As a consequence ipsilateral peripheral vascularity is attenuated and pulmonary artery becomes small. Usually left side of unknown reasons is commonly involved however bilateral involved can also be seen [11].

On imaging Macleod syndrome is characterized by unilateral loss of lung volume with hyperlucency on chest skiagram, reduction in vascularity on CT scan of the chest, hypoplastic ipsilateral pulmonary artery associated with air trapping on expiratory scans [128]. In our case initially expiratory scans were not done however after strong diagnostic possibility of this syndrome expiratory chest X-ray was done which revealed air trapping there by confirming our diagnosis. Radionuclide scans are corroborative and reveals unilateral loss of perfusion on Technetium 99c lung scan and air trapping on ventilation scan using xenon [124].

Swyer-James-Macleod syndrome has a diverse manifestations in adults, may present with recurrent pulmonary infections or hemoptysis due to associated bronchiectasis, and mild respiratory symptoms. The course is generally a stable one [127812].

The differential diagnosis of the chest X-ray includes proximal interruption of pulmonary artery, the hypogenetic lung syndrome and pulmonary artery obstruction due to embolism; however none of these conditions exhibits air trapping [124]

In conclusion if in a symptomatic patient X-ray chest shows unilateral hyper transradiancy with air trapping on expiratory films, this entity should be suspected and a CE- HRCT chest should always be done to diagnose this syndrome and to detect associated complications like bronchiectasis.

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