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SWYER JAMES MACLEOD SYNDROME "AN INCIDENTAL FINDING IN A MIDDLE AGED WOMEN WITH A UNILATERAL HYPERLUCENT LUNG"

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ABSTRACT
Swyer-James Macleod syndrome also k/a Bret syndrome is an exceptional form of post infectious obliterative bronchiolitis usually thought to be secondary to Viral infections. It is consequence of insult abide by which it results hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion .It is seen in association with 3.8% patients with bronchiolitis obliterans. The usual age of presentation is early childhood or sometimes adulthood. Patients are usually asymptomatic. The most concerning long-term effects of this syndrome is recurrent lung infections. Most patients have a unilateral involvement of the left lung primarily. In some patients with Swyer-James-MacLeod syndrome, bronchiectasis may develop proximal to bronchiolitis obliterans and is likely a sequela of chronic infection

KEYWORDS: Obliterative bronchiolitis, Lung hypoplasia, Lung hypoperfusion, Lung agenesis.

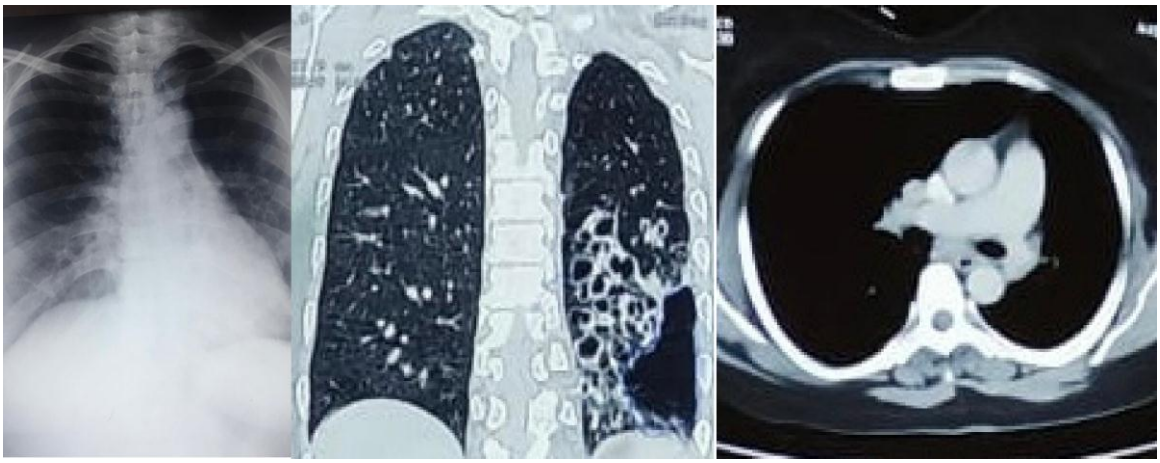
BACKGROUND
Swyer James syndrome is a unique form of post infectious obliterative bronchiolitis. The usual precipitating factor is secondary to acquired viral and bacterial infection. It is characterized by hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion. The usual age of presentation is early childhood or sometimes adulthood. Most patients have a unilateral involvement of the left lung primarily. The characteristic radiographic findings include a unilateral hyperlucent lung along with decreased bronchovascular markings, small hilar shadow and slight displacement of mediastinum to the affected side. Bronchiectasis is seen in 30% of patients (cystic or saccular). Individuals with SJS who have saccular bronchiectasis usually have more severe exacerbation.

CASE REPORT
28 years female non-smoker presented to OPD with complains of shortness of breath for 10 years increased since last 10 days which was gradual onset, progressive, MMRC-II was associated with seasonal variation. She had history of chronic recurrent cough for 10 years (initially dry later productive for last few years). She had history of fever for 10 days. She had history of recurrent pulmonary infection for which she was repeatedly admitted to hospital. She also had history of MDI use for 8 years, ATT intake for pulmonary TB which was microbiologically confirmed for 6 months. She denied any complains of hemoptysis, orthopnea, PND. No H/o HTN, DM, Hypothyroidism. No h/o surgery

On examination: Patient was febrile and tachypneic. On auscultationB/L diffuse wheeze with fine crepts on left infraxillary area was present. Her systemic examination was unremarkable. Laboratory findings revealed normal routine count and a differential count. The renal and liver parameters were within normal range.The sputum routine microscopy revealed greyish white, 88% Neutrophils, 9% lymphocyte and 3% monocytes. **Gram stain:** gram positive and gram negative bacilli. **Sputum c/s:** Pseudomonas, Serum Total IGE -1752 (0-113); IGE specific for Aspergillus – 0.11 (<0.1);IGG specific for Aspergillus – 9.56(<27).

Digital CXR, **CECT thorax**

Small hyperlucent left lung with mediastinal shift towards left .Cystic bronchiectasis in the lingula and left lower lobe with 'bunch of grapes' appeareance. Left pulmonary artery is narrow measuring 10.4 mm, right pulmonary artery 21 mm diameter. Emphysematous bulla in left lower lobe.



Management

Patient was managed with antibiotics (dual antibiotics)- A macrolide and a broad spectrum antibiotic.After the reports of culture in which Pseudomonas was isolated the antibiotics was changed according to culture sensitivity reports. Patient was given antibiotics as long as 3 months with repeat cultures on day 14.

Low dose inhaled corticosteroids and inhaled bronchodilators, mucolytics, chest physiotherapy was done. During discharge pneumococcal and influenza vaccination was given.

Nutrition and supportive therapy

i. Protein rich diet and sufficient vitamins, minerals and antioxidants
ii. Immune modulators and indigenous herbal preparations to boost immunity of patient may be helpful

DISCUSSION

In this case the first impression was consistent with bronchiectasis in left lower lobe.As she was giving history of recurrent infection and use of inhaler so the first thing we worked up in the line of ABPA as she was giving history that mimics Asthma and along with bronchiectatic changes in CXR we worked up in that line as well. We thought Tuberculosis was less likely because there was lower lobe involvement and also she complained of recurrent infections. Since there was a past history of ATT intake we sent the sample for gene expert and sputum culture. Finally radiological hyperlucency in HRCT in left lung with mediastinal shift towards left, with narrow left pulmonary artery measuring 10.4 mm, right pulmonary artery 21 mm diameter was characteristically associated with SJMS along with saccular bronchiectasis. Literature have shown that patient with saccular bronchiectasis have more severe infection and exacerbation. Sputum culture showed that she had secondary bacterial infection with Pseudomonas. A ventilation-perfusion lung scanning could also be performed but false-positives could appear in the presence of any disorder involving distal airway

obstruction (such as, bronchiolitis obliterans, asthma or congenital lobar emphysema).

CONCLUSION

The present case highlights that a chest X-ray may underestimate the prevalence of the SJMS syndrome. Irrespective of numerous chest radiographic examinations, SJMS was not diagnosed until another complementary imaging was performed. The reason for reporting this case was because of fact that few cases have been reported worldwide of adults presenting with SJMS. In context of our country due to inaccessibility of health services due to socio-economic and cultural strains. Delay in early diagnosis results in complication and more severity of exacerbations.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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