RESEARCH PAPER | Medical Science | Volume : 6 | Issue : 2 | FEBRUARY 2016 | ISSN - 2249-555X

Swyer-James Syndrome- a Rare Case Report

KEYWORDS: Swyer-James syndrome, Swyer-James –MacLeod syndrome.

Dr Uddhav S.Khaire	Dr Vimlesh R Pandey
Asst. Pro, Department of Medicine, GMCH, Aurangabad	JRIII Department of Medicine, GMCH, Aurangabad

Dr Shradha S. Runwal	Dr Pranay G. Jambhulkar
JRII Department of Medicine, GMCH, Aurangabad	JRI Department of Medicine, GMCH, Aurangabad

ABSTRACT A 79 year male patient presented to us with a history of fever, cough with expectoration, breathlessness on exertion increased since 10 days. After thorough examination and investigations including chest X-ray and CECT chest, a diagnosis of Swyer-James syndrome was made.

Introduction:
For the first time, this syndrome was defined by Swyer and James in 1953 in a 6-year-old boy with unilateral emphysema treated with pneumonectomy. A year later, Macleod reported a series of 9 cases with unilateral hyperlucency. From this date on, the disease was defined as Swyer-James-Macleod syndrome (SJMS).[1]Our case presented with fever, cough, and breathlessness since 10 days. On further investigating with X-ray chest and CECT thorax a diagnosis of Swyer-James syndrome was made.

Case history:
A 70 year elderly male patient presented to us with a history of fever, cough with expectoration, breathlessness on exertion increased since 10 days. Patient had similar complaints in past for which he was on bronchodilator therapy since 10 years. Patient was a chronic tobacco chewer. Patient did not have a history of tuberculosis in past. Sputum was negative for acid fast bacilli .On blood investigations, complete blood counts, renal and liver function tests were within normal limits. X-ray chest was suggestive of mediastinal shift to left, cystic bronchiectasis and hypoplastic left lung field (as shown in Fig.1). So CECT chest was done (as shown in Fig.2) which revealed volume loss in posterior segment of left upper lobe and left lower lobe with mediastinal shift towards left, cystic bronchiectatic changes in entire left lung and lower lobe of right lung, left pulmonary artery showed abrupt narrowing suggestive of hypoplastic left pulmonary artery with few collaterals, all the findings were highly suggestive of a diagnosis of Swyer-James syndrome. As patient was symptomatically better, patient was discharged on bronchodilator therapy.

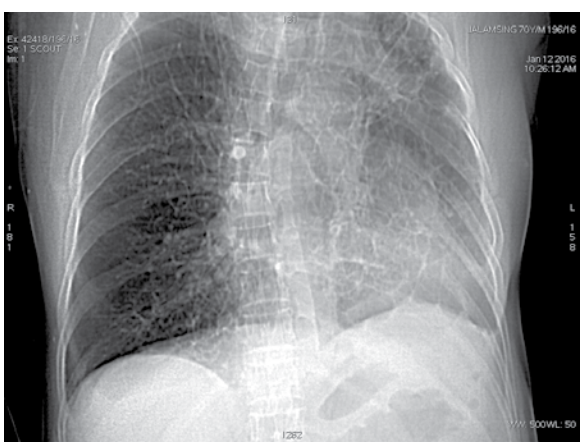

Fig.1

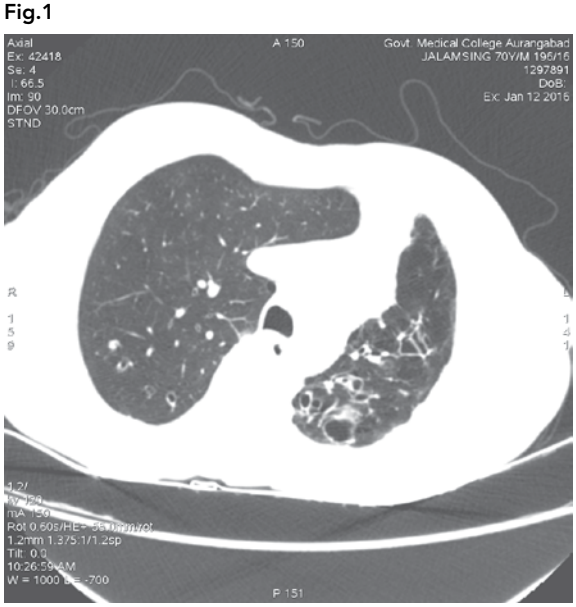

Fig.2

Volume : 6 | Issue : 2 | FEBRUARY 2016 | ISSN - 2249-555X

Conclusion:
An elderly male patient presented to us with acute febrile illness, and on investigating further, as CECT thorax was suggestive of left pulmonary artery hypoplasia with hypoplasia of posterior segment of left upper lobe and left lower lobe of lung , a diagnosis of a very rare entity Swyer-James syndrome was made, hence presenting this case report.

Discussion:
Swyer-James-MacLeod syndrome or unilateral hyperlucent lung syndrome is a rare entity associated with postinfectious bronchiolitis obliterans occurring in childhood. It is characterized by hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion.[2]

Typically, this disorder is diagnosed in childhood after an evaluation for recurrent respiratory infections but sometimes patients who have little or no sequelae bronchiectasis have minor symptoms or are asymptomatic and may, therefore, miss their diagnosis until adulthood.[3]

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. [3]

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.[3]

Bronchiolitis obliterans results with inflammation of the respiratory bronchiole wall, fibrosis, and luminal narrowing. The fibrosis, in the intra-alveolar septa results with obliteration of the pulmonary capillary bed resulting in blood flow decrease in the main pulmonary artery segments. As a result, hypoplastic arterial formation occurs. The decrease of the compensatory perfusion in the peripheral respiratory airways develop. These pathophysiologic changes lead to air trapping and hypoperfusion in the affected segment, thus creating radiographic hyperlucent or translucent findings .[1]

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.[2]

Characteristically in CT angiography, pulmonary artery hypoplasia/agenesis and decrease of pulmonary artery calibration can be detected, thus decrease of pulmonary blood supply can be detected. Other characteristic findings are pathologic perfusion changes at the affected lung, heterogeneous patch like air trapping, and atelectasis of healthy pulmonary areas due to compression.[1]

SJMS treatment includes the early control of lung infections as well as influenza and pneumococcal vaccinations .No specific morbid-mortality studies with SJMS have been done.[4]

REFERENCE 1. Hatice Kaplanoglu, Veysel Kaplanoglu, Ugur Toprak, Alper Dilli, and Baki Hekimoglu , A Case of Swyer-James-Macleod Syndrome Associated with Middle Lobe Hypoplasia and Arteriovenous Malformation. Case Reports in Medicine Volume 2012 (2012); Article ID 959153: 4 pages 2. Carlos Capela, Paulo Gouveia, Marco Sousa, and Maria J Regadas, Adult diagnosis of Swyer-James-MacLeod syndrome: a case report. J Med Case Reports. 2011; 5: 2. 3. Carlos Capela12, Paulo Gouveia, Marco Sousa and Maria J Regadas, Adult diagnosis of Swyer-James-MacLeod syndrome: a case report. Journal of Medical Case Reports 2011; 5:2 doi: 10.1186/1752-1947-5-2 4. Carlos Capela; Paulo Gouveia; Marco Sousa; Maria J Regadas, Adult Diagnosis of Swyer-James-MacLeod Syndrome: A Case Report.J Med Case Reports. 2011; 5: (1)