EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 8
Issue: 10
83-85
Year: 2021

SWYER-JAMES-MACLEOD SYNDROME IN A PATIENT WITH OCULAR MYASTHENIA GRAVIS AND MEDIASTINAL THYMOMA: RADIOLOGICAL REVIEW OF UNUSUAL ASSOCIATION

[1*]Dr. Akshay Sharma, [2]Dr Deeksha Sharma, [3]Dr. Alaipreet Kaur, [4]Dr. Shefali Singla, [5]Dr. Archita Gupta and [6]Dr. Ankita Jindal

[1]MD Radiodiagnosis, Civil Hospital Shahpur, Himachal Pradesh, India.
[2]Department of Pharmacology, Dr. RPGMC Kangra at Tanda, HP India.
[3]Department of Radiology, Dr. RPGMC Kangra at Tanda, HP India.
[4]Dr. RPGMC Kangra at Tanda, HP India.
[5]Department of Radiology, Dr. RPGMC Kangra at Tanda, HP India.
[6]Department of Radiology, Dr. RPGMC Kangra at Tanda, HP India.

***Corresponding Author: Dr. Akshay Sharma**
MD Radiodiagnosis, Civil Hospital Shahpur, Himachal Pradesh, India.

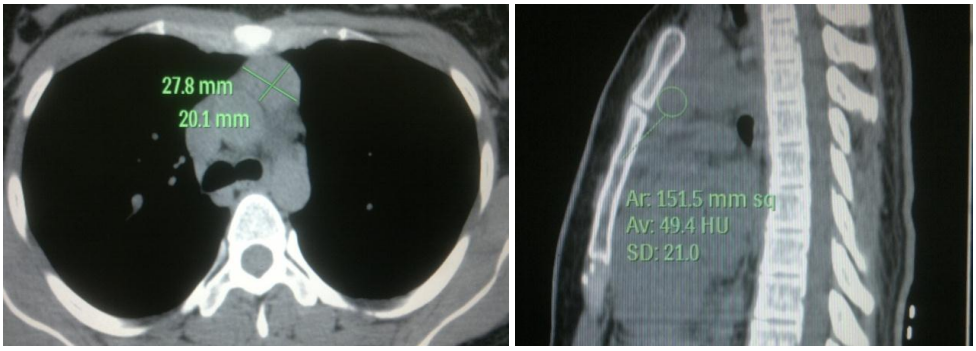
Article Received on 26/07/2021 **Article Revised on 16/08/2021** **Article Accepted on 05/09/2021**

ABSTRACT
Swyer-James-MacLeod syndrome is post infective constrictive bronchiolitis which causes air way obstruction. The patient can be asymptomatic and can also present with complaints like shortness of breath, cough, repeated chest infections. Radiological investigations play an important role in accurate diagnosis of this syndrome and help in starting timely treatment. A 30 year old female patient with ocular myasthenia gravis, referred for CECT thorax to rule out thymoma.

KEYWORDS: Swyer-James-MacLeod syndrome (SJMS), Hyperlucent lung syndrome, Computed tomography (CT).

INTRODUCTION
Swyer-James Syndrome or Swyer-James-Macleod Syndrome is also known as unilateral hyperlucent lung syndrome. In 1953, a physician Paul Robert Swyer and a radiologist George James in Canada described a case of unilateral pulmonary emphysema in a 6 year old boy with multiple episodes of bronchopneumonia and bronchitis.[1] In 1954 , William Mathieson Macleod in England who was a respiratory physician gave detailed description of 9 cases with abnormal lucency of one lung.[2] Based on their names, this syndrome was called as Swyer-James-Macleod Syndrome . This syndrome is associated with post infective bronchiolitis obliterans.

CASE
29 years old female patient with diagnosed ocular myasthenia gravis was referred for contrast enhanced CT (CECT) thorax to rule out thymoma. She had history of multiple episodes of bronchopneumonia in childhood. She had no respiratory complaints at the time of CT scan. Patient was non smoker. No history of ATT intake. No history of fever, weight loss, haemoptysis. CECT thorax was done which showed homogenous soft tissue attenuation mass in retro sternal region showing mild homogenous enhancement. No internal calcification, fat attenuation areas seen. Overlying bone was normal. Surrounding fat planes were maintained. No significant mediastinal lymphadenopathy seen. Based on these findings this retrosternal mass was diagnosed as thymoma. There was paucity of vessels in left upper zone with no proximal interruption / filling defect .Right and left pulmonary arteries were normal. On lung window there was presence of geographic hypoattenuating area in left upper lobe involving apical and anterior segment with vascular paucity in affected area. Smaller and less number of vessels were seen in the affected area .No large airway agenesis seen. Large airways were patent with no intraluminal obstruction. No bronchial wall thickening and dilated bronchi seen. No architectural distortion, mediastinal shift, lobar collapse, bronchiectatic changes seen. Based on these findings diagnosis of Swyer-James-Macleod Syndrome was made.



A b

Figure 1 (a, b): Non contrast CT Thorax axial (a), sagittal(b) images showing homogenous soft tissue attenuation mass in retro sternal region. No internal calcification, fat attenuation areas seen. Overlying bone was normal. Surrounding fat planes were maintained.

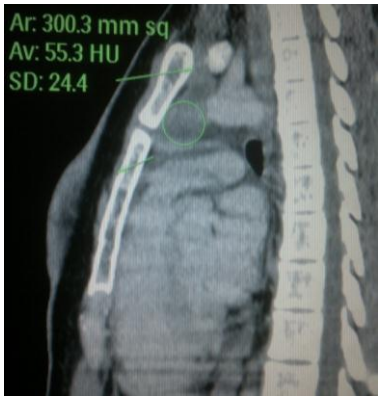


Figure 2: Sagittal CECT Thorax showing mild homogenous enhancement in the retrosternal mass.

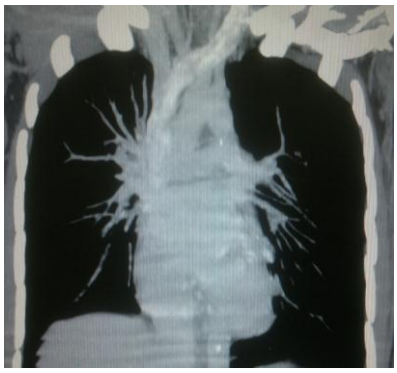
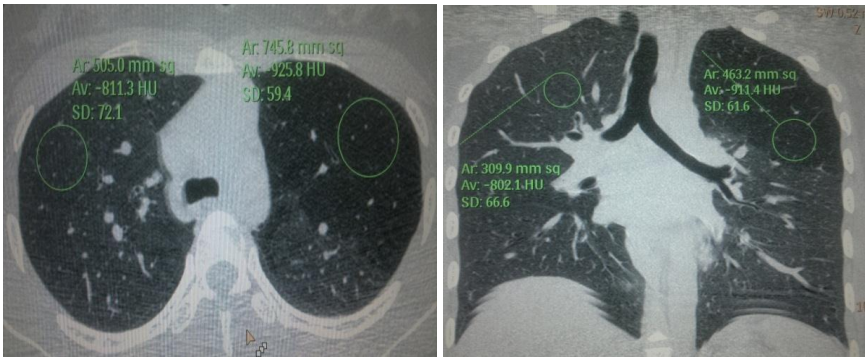


Figure3: Coronal CECT Thorax MIP image shows paucity of vessels in left upper zone.



A b

Figure 4: Lung window CT Thorax axial (a) and coronal (b) images showing geographic hypoattenuating area in left upper lobe involving apical and anterior segment with vascular paucity in affected area. No bronchiectatic changes seen.

DISCUSSION

Swyer-James-Macleod Syndrome occurs as delayed complication of infectious bronchiolitis in early childhood caused by organisms like Mycoplasma pneumonia, adenovirus, influenza A, mycobacteria, paramyxovirus, Bordetella pertussis and measles.[3] As a result of inflammatory response to the infection, damage to the terminal and respiratory bronchioles occurs. There occurs peribronchiolar fibrosis and luminal obliteration of the bronchioles which impairs development of the alveolar bud leading to incomplete development of alveoli and involved segment of lung.[4] Long term sequalae include destruction of lung parenchyma, bronchiectasis leading to recurrent lung infections, lung abscesses, spontaneous pneumothorax. [5,6] Inflammation also causes damage to lung vasculature leading to decreased peripheral lung vascularisation.

There is definite association of myasthenia with thymus hyperfunctioning because 70% of myasthenia patients have thymic hyperplasia and upto 15% patients have thymoma.[7] On review of literature no definite association of myasthenia gravis and thymoma with SJMS was found.

On Chest X-ray , the most common finding in SJWS is hyperlucent one lobe or lung , however it can be bilateral. This hyperlucency occurs due to air trapping. The air enter the air spaces by collateral air drift but cannot exit because of the bronchiolar obstruction (because of post infectious bronchiolitis obliterans).[8] In Swyer-James-Macleod Syndrome we have to exclude others causes of obstruction like tumour and foreign body.[8] There is presence of sparse ,attenuated vessels in the involved part of lung. Both the above findings become more prominent on expiratory films. Also the volume of the hyperlucent lung do not change on expiration.

Computed Tomography of thorax plays an important role in estimation of extent and distribution of SJMS. Also

CT is more sensitive in detecting hyperlucent lung regions and their various differential diagnosis.[9] CT shows characteristic findings of hyperlucent lung with attenuated and sparsely distributed vessels. Air trapping is seen on expiratory scan. CT is more sensitive in detecting and characterising bronchiectasis, lung abscesses, mosaic perfusion.

The role of CT is very important because in many cases SJM syndrome is confused with other lung diseases like COPD, asthma due to similar clinical presentations and the patient gets inappropriate treatment for long time until accurate diagnosis is made with the help of computed tomography.

In this way radiological investigations help in accurate diagnosis of SJMS so that timely treatment can be started. Treatment options are mainly symptomatic with active management of recurrent pulmonary infections.[10]

REFERENCES

1. Swyer PR, James GC. A case of unilateral pulmonary emphysema. Thorax, 1953 Jun; 8(2): 133-6.
2. Macleod WM. Abnormal transradiancy of one lung. Thorax, 1954 Jun; 9(2): 147-53.
3. Damle NA, Mishra R, KaurWadhwa J. Classical imaging triad in a very young child with Swyer-James syndrome. Nucl Med Mol Imaging, 2012; 46(2): 115–8.
4. Yekeler E. A rare case of Swyer-James Macleod syndrome and a new clinical presentation, acquired lobar emphysema. Ann Thorac Surg., 2012; 93(5): e123–5.
5. Wang JY, Chen KY, Chang YC, Chang YL, Lee LN. Swyer-James syndrome complicated by lung abscess. J Formos Med Assoc., 2000; 99(3): 252-256.
6. Soni R, Barnes D. Macleod's syndrome presenting with spontaneous pneumothorax. Respirology, 1999; 4(3): 275-277.
7. Sommer N, Melms A, Weller M, Dichgans J. Ocular myasthenia gravis. A critical review of clinical and pathophysiologicalaspects. Doc Ophthalmol., 1993; 84: 309–33.
8. Becroft DMO. Bronchiolitis obliterans, bronchiectasis, and other sequelae of adenovirus type 21 infection in young children. J Clin Pathol., 1971; 24: 72-82.
9. Moore AD, Godwin JD, Dietrich PA et-al. Swyer-James syndrome: CT findings in eight patients. AJR Am J Roentgenol., 1992; 158(6): 1211-5.
10. Tutar O, Tekcan DE, Samanci C, Bas A. Adult diagnosis of Swyer James-Macleod syndrome. BMJ Case Rep., 2012. doi:10.1136/bcr2012-007349.