Case report

Swyer-James-MacLeod Syndrome: a rare case

Singal KK[1], Bhatti KP[2], Gupta A[3], Gupta N[4], Samra R[5],Gautam S[6]

Abstract

This case report describes a patient with Swyer-James-MacLeod Syndrome (SJMS) in an adult female diagnosed on the basis of findings on X-ray chest, high resolution CT (HRCT) of chest. This rare syndrome is considered to be an acquired disease due to repeated pneumonias in early childhood.

Keywords: *Swyer-James syndrome; chest X-ray; high resolution computed tomography; bronchiectasis*

Bangladesh Journal of Medical Science Vol. 14 No. 04 October'15. Page: 402-404
DOI: http://dx.doi.org/10.3329/bjms.v14i4.20407

Introduction

Swyer-James syndrome also known as MacLeod syndrome, first described in 1953, is an uncommon disease characterized by hyperlucency of one lung,lobe or part of lobe due to pulmonary vascular abnormalities and alveolar overdistension[1]. Emphysema, bronchiectasis or bronchiolitis obliterans are the cardinal pathological features[2]. The syndrome is of unknown etiology although repeated episodes of viral bronchiolitis or viral pneumonias are the speculated cause[1]. Patients usually present as adults and prior history of childhood infections is not obtained in most cases[3]. This report describes a case of Swyer-James syndrome diagnosed in an adult female.

Case report

A 32 years old female presented with history of gradually progressive dyspnea, grade III, for last six months. There was history of associated cough, sputum (occasional episode of blood),fever , anorexia and weight loss. She had no past history of tuberculosis, asthma, childhood pneumonias, whooping cough or measles. She had received complete coverage of Expanded Program of Immunization (EPI) in early childhood. She was a non-smoker and house wife by occupation .There was no history of exposure to pets, farm animals, organic or inorganic dust. She was married for the last seven years with no complication during her two past pregnancy. On examination, she had respiratory rate of 22/minute with tracheal shift to the right, resonant percussion note and added breath sounds on the left side of chest i.e diffuse crackles on left upper and lower zone. On investigations, Montoux's test, C-reactive protein (CRP) and sputum for AFB were negative. ESR was 30 mm at the end of first hour. Analysis of arterial blood gases showed pH of 7.42 with PaO2 of 144 mmHg, PaCO2 of 26.2 mmHg and arterial oxygen saturation of 99.2%. Spirometry showed restrictive ventilatory defect. X-ray chest PA film showed marked hyperlucency of left lung collapse of left lower lobe and mediastinal shift to the left (Figure 1). HRCT of chest showed loss of volume of left lung with partial collapse of left lower lobe with extensive cystic bronchiectasis with air trapping and relatively small sized left pulmonary artery (Figure 2). She was given supportive treatment including bronchodilators, antibiotic courses as and when required and was advised to have pneumococcal and influenza vaccines on regular basis as per schedule. Patient showed symptomatic improvement with treatment and was advised to have regular follow-up with her physician.

1. Dr. Kiran Kumar Singal, Associate Professor , Department of Medicine
2. Dr. Karun Puran Bhatti, Assistant Professor, Department of Medicine
3. Dr. Abhinav Gupta , Resident , Department of Medicine
4. Dr. Nitin Gupta, Resident, Department of Medicine
5. Dr. Ravneet Samra, Resident, Department of Medicine
6. Dr. Sumit Gautam, Resident, Department of Medicine
M.M.Institute of Medical Sciences & Research, Mullana,Ambala(133203)India

Corresponds to: Dr. Kiran Kumar Singal, Department of Medicine ,M.M.Institute of Medical Sciences & Research,Mullana,Ambala(133203) India.Email: drkiranksingal@yahoo.co.in

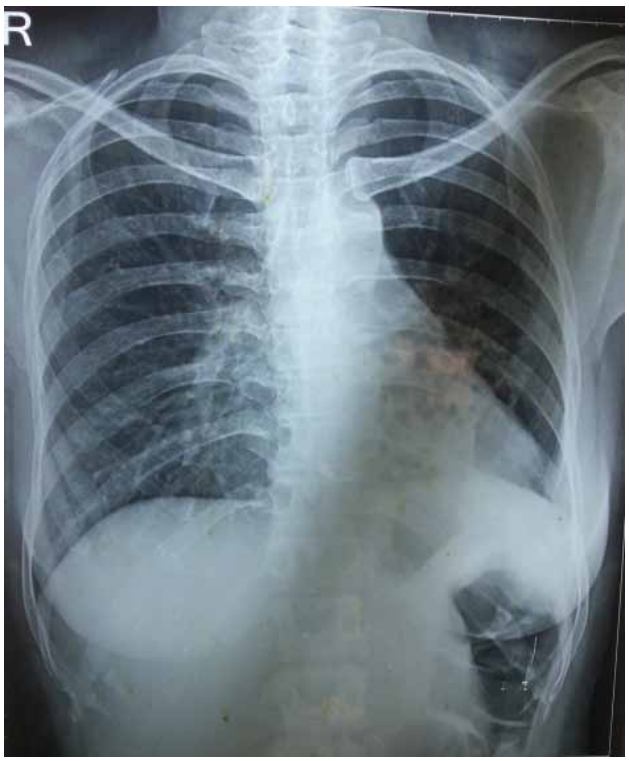

Figure 1: X-ray chest showig hyperventilated right lung, collapsed lower lobe left lung with multiple air trapped area underneath cardiac shadow and mediastinal shift to the left.

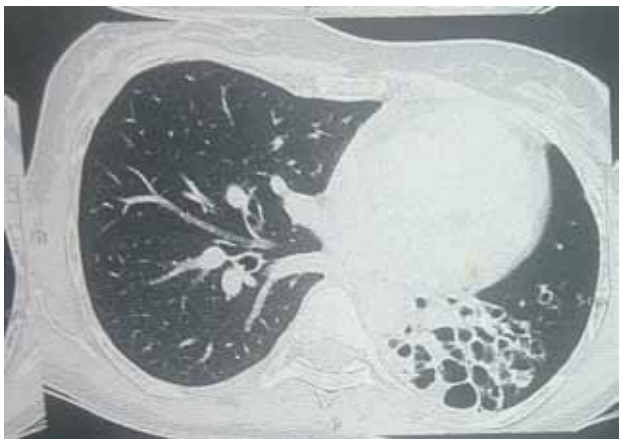

Figure 2: HRCT of chest showing loss of volume of left lung with partial collapse of left lower lobe with extensive cystic bronc hiectasis with air trapping and relatively small sized left pulmonary artery.

Discussion

Swyer-James syndrome is an uncommon cause of localized hyper-transradiancy of lung. It is characterized by unilateral hyperlucency of lung on X-ray chest, unilateral reduction in vascularity on chest CT and unilateral loss of perfusion on lung scan[4,5]. Due to unknown factors, it usually involves the left lung[6]. It is a form of obliterative bronchiolitis with concomitant vasculitis following injury to immature lungs during the first eight years of life. It usually follows infections with organisms like adenovirus, measles or pertussis. Both small bronchi and bronchioles are affected and the lung with abnormal airways remains inflated by collateral air drift[7]. This damage during the early childhood prevents normal development of the alveolar ducts. Airways develop submucosal fibrosis leading to luminal irregularity and occlusion. Pulmonary vasculature is hypoplastic while the lung distal to diseased bronchioles become hyperinflated and sometimes pan acinar emphysematous changes develop[7]. Patients usually present with gradually progressive dyspnea and repeated chest infections because of associated bronchiectasis, although this patient denied any history of cough with copious purulent sputum[2]. Spirometery in these patients usually show restrictive defect as in this case, though a significant proportion of patients can have obstructive pattern due to associated bronchiolitis and bronchiectasis[7]. The disease as assessed radiologically is predominantly unilateral so other causes of hyperlucent lung like pneumothorax, asymmetric emphysema, congenital lobar emphysema(CLE), pulmonary artery hypoplasia, pulmonary embolism and bronchial obstruction due to foreign body or mucus plugs should be ruled out[1]. Hyperlucency in this case is due to reduced perfusion and air trapping. Pulmonary vessels are reduced on the affected side but lung volumes are only slightly decreased. Ipsilateral air trapping is a key finding[7]. The characteristic radiographic findings in these patients include unilateral hyperlucent lung along with decreased broncho vascular markings, a small hilar shadow and slight displacement of the mediastinum to the affected side. In this case, mediastinum was shifted to the contralateral side because of the long-standing nature of the illness and severe emphysematous changes in the left lung[10]. CT scan findings of Swyer-James syndrome include patchy areas of low attenuation and hypovascular areas interspersed with areas of normal attenuation. Air trapping is confirmed on expiratory scans. Other changes on CT may include bronchiectasis, bronchiolectasis, atelectasis and scarring[8,9]. Bronchiectasis, though not a universal finding, evident in only 30% of patients, affects the clinical manifestations and prognosis of the disease. Patients who had saccular bronchiectasis usually have more severe exacerbations than those who do not have bronchiectasis on HRCT. In this case, bronchiectasis

was identified on HRCT chest and this may explain her apparent presence of symptoms for a longtime till she developed gradually
progressive dyspnea. It cannot, though reliably differentiate congenital from acquired causes of hypoplastic pulmonary vasculature. Pulmonary angiography was not performed in this patient for this reason. Treatment is largely supportive with early control of super-added infections along with influenza and pneumococcal vaccination[1]. Bronchodilators may help, especially if the spirometery shows obstructive defect. Long-term oxygen therapy is required in cases with advanced disease and respiratory failure. Surgery in the form of lobectomy or pneumonectomy can be offered to those who had severe symptoms despite optimal therapy. Prognosis largely depends on presence of associated bronchiectasis[4,10]. This patient had emphysema and have bronchiectasis, so prognosis in this individual case may be fair. Regular follow-up and ancillary care is required.

References

1. Tortajada M, Gracia M, Gracia E, Hernandez R. Diagnostic Journal of the College of Physicians and Surgeons Pakistan 2008, Vol. 18 (3): 190-192 **191** Swyer-James-MacLeod syndrome considerations in unilateral hyperlucency of the lung (Swyer- James-MacLeod syndrome). *Allergol Immunopathol* (Madr) 2004;**32**: 265-70.
2. Marchevesky AM, Guintu R, Koss M, Fuller C, Houck W, McKenna RJ. Swyer-James-MacLeod syndrome with placental transmogrification of the lung: case report and review of the

literature. *Arch Pathol Lab Med* 2005;**129**:686-9.

3. Mera A, Ichikado K, Wada M, Miyajima M, Suga M. A case of Swyer-James syndrome with interesting chest radiographs. *Nihon Kokyuki Gakkai Zasshi* 2006; **44**:354-8.
4. Quaedvlieg V, Duysinx B, Ghaye B, Bartsch P. Clinical case of the month. MacLeod syndrome. *Rev Med Liege* 2004; **59**:691-4.
5. Chan PW, DeBruyne JA, Goh AY, Muridan R. Swyer-James-MacLeod syndrome. *Med J Malaysia* 1999; **54**:520-2.
6. Abba AA, Al-Mobeireek AF. Clinical spectrum of Swyer-James-MacLeod syndrome in adults. *Saudi Med J* 2003; **24**:195-8.
7. Collins J, Stern EJ. Unilateral hyperlucent lung. In: Collins J, Stern EJ, (edi). Chest radiology: the essentials. Philadelphia:*Lippincott Williams & Wilkins*;1999.p.207-8.
8. Moore AD, Godwin JD, Dietrich PA, Verschakelen JA,Henderson WR Jr. Swyer-James syndrome: CT findings in eight patients. *Am J Roentgenol* 1992;**158**:1211-5.
9. Marti-Bonmati L, Ruiz Perales F, Catala F, Mata JM, Calonge E. CT findings in Swyer-James syndrome. *Radiology* 1989; **172**:477-80.
10. Gopinath A, Strigun D, Banyopadhyay T. Swyer-James syndrome. *Conn Med* 2005; **69**:325-7