ORIGINAL RESEARCH PAPER
Volume - 11 | Issue - 02 | February - 2022 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

SWYER-JAMES-MACLEOD SYNDROME (SJMS) WITH PULMONARY HYPERTENSION: AN UNUSUAL CASE REPORT WITH REVIEW OF LITERATURE

Pulmonary Medicine

Dr Mohd Imran Shamsi* Senior Resident, Department of Pulmonary Medicine And Critical Care, Metro Centre for Respiratory Diseases(MCRD), Metro Multispeciality Hospital, Noida, Uttar Pradesh, India *Corresponding Author

Dr Rohit Vadala Junior Consultant, Department of Pulmonary Medicine And Critical Care, Metro Centre for Respiratory Diseases(MCRD), Metro Multispeciality Hospital, Noida, Uttar Pradesh, India.

Dr Deepak Talwar Director And Head, Department of Pulmonary Medicine And Critical Care, Metro Centre for Respiratory Diseases(MCRD), Metro Multispeciality Hospital, Noida, Uttar Pradesh, India.

ABSTRACT

Swyer-James-Macleod syndrome (SJMS) is an uncommon disorder of the lungs resulting in unilateral hyperlucent lungs and decreased broncho-vascular markings. It is typically diagnosed during childhood, although occasionally some patients are diagnosed in adulthood as an incidental finding. Pulmonary hypertension complicating this syndrome is very rare which poses a diagnostic dilemma and a challenge to manage such patients. We describe this unique case report which was initially misdiagnosed as uncontrolled bronchial asthma and later turned out to be Swyer-James syndrome associated with pulmonary hypertension.

KEYWORDS

Swyer-James-Macleod syndrome, pulmonary hypertension, hyperlucent, bronchial asthma.

Case Report

A 12-year-old school going boy, resident of Uttar Pradesh, India with no prior co-morbidities; was referred to our facility with increasing progressive breathlessness, modified medical research council (mMRC) grade 1 to grade 3 along with recurrent streaky haemoptysis for the past 6 months. He had a past history of recurrent lower respiratory tract infections since birth and he was treated with inhaled bronchodilators by local medical practitioner with not much symptomatic relief. The patient was previously admitted at elsewhere hospital 6 months ago with similar presentation and was managed conservatively. The patient denied smoking or drug use. He also denied headaches, dizziness, syncope, paroxysmal nocturnal dyspnoea or pedal oedema. His personal and family history was unremarkable.

On initial evaluation, the patient was afebrile. He was haemodynamically stable with blood pressure of 110/60 mm Hg. He was tachypneic and tachycardia was noted. Pulse oximetry saturation was 92% on room air. His general physical examination showed no pallor, icterus, clubbing or pedal oedema. JVP was not raised and neck veins were not engorged. Cardiovascular system examination revealed pan systolic murmur over lower left sternal border. Respiratory system examination revealed decreased breath sounds over both bases.

His laboratory parameters revealed haemoglobin of 14gm/dl, total leukocyte count (TLC) of 7000/mm^3, with normal platelets counts. Renal and liver function tests were normal, sputum for pyogenic culture was negative and GeneXpert for mycobacterium tuberculosis was not detected. Erythrocyte sedimentation rate (ESR) was normal and viral markers for hepatitis B & HIV were negative. Electrocardiogram showed normal sinus rhythm with tachycardia. Arterial blood gases revealed mild hypoxemia. D-dimer level was within normal limits and N-terminal brain natriuretic peptide value was 90pg/ml.

2D echocardiogram (ECHO) revealed normal left ventricular ejection fraction (EF) of 55%, dilated right atrium and right ventricle along with straightening of interventricular septum with moderate right ventricular systolic dysfunction suggesting right heart strain. Trivial tricuspid regurgitation was noted along with dilated main pulmonary artery. ECHO findings showed severe pulmonary hypertension with estimated mean pulmonary artery pressure (mPAP) of 55mmHg.

Chest X-ray showed hyper inflated right lung with decreased left hemithorax volume. Contrast enhanced computed tomography (CECT) of the chest with pulmonary angiogram was done which showed reduced left hemithorax volume with hyperinflation of right lung. Left lung was more lucent with reduced broncho-vascular markings. Both main pulmonary artery (MPA) and right pulmonary artery (RPA) were enlarged. Atresia of the left pulmonary artery was noted beyond left hilum. There was no evidence pulmonary thromboembolism. CT findings were consistent with Swyer-James-Macleod syndrome.

Pulmonary function testing showed pre-bronchodilator and post-bronchodilator forced expiratory volume in 1 second (FEV_1) of 0.94L(45% predicted) and 1.10L(53% predicted) respectively, pre-bronchodilator and post-bronchodilator forced vital capacity (FVC) of 1.53L(62% predicted) and 1.84L(74% predicted) respectively, FEV_1/FVC of 59.74. Total lung capacity (TLC) of 3.07L (91% predicted) and diffusion capacity was 10.38ml/min/mmHg (56%). 6-minute walk test (MWT) performed showed significant oxygen desaturation from 92% to 84% on walking distance of 336 meters

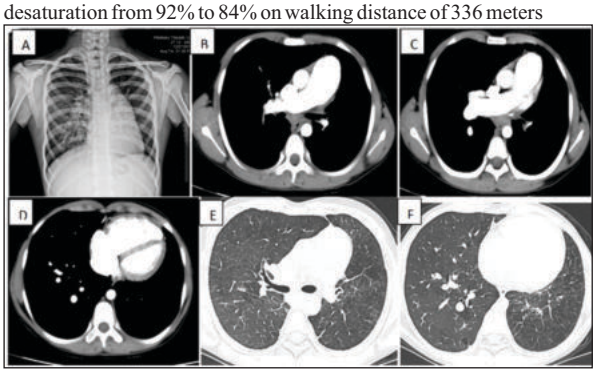

Figure 1: A- Chest X-ray showing left lung hyperlucency with decreased vascular markings; **B, C, D-** Axial sections of CTPA showing atresia of left pulmonary artery (LPA) with decreased broncho-vascular marking on left side; **E, F-** HRCT chest showing hyperlucent left lung along with decreased vascular markings and hyperinflation of right lung with herniation into left side

Patient was managed with antibiotics along with oxygen therapy, inhaled corticosteroids and bronchodilators. Cardiac angiography with right heart catheterization was advised to patient; however, it was deferred by the patient's family. Patient was later given pneumococcal and influenza vaccines. Paediatric cardiologist opinion was taken and the patient was discharged on anti-pulmonary hypertension (PH) therapy; phosphodiesterase- 5 inhibitors (PDE-5), sildenafil 20mg

once a day titrating it to 20mg thrice a day after monitoring systemic blood pressure. Subsequently, selective endothelin receptor- A (ETA) antagonist, ambrisentan 5mg was added at 2 weeks follow up visit in view of persistent exertional breathlessness. Exercise capacity by 6 MWT increased from 336 meters to 370 meters with baseline oxygen saturation of 95% and nadir of 90% on dual anti-PH therapy. Patient was later referred for combined heart and lung transplantation in view of persistent clinical symptoms and right ventricular dysfunction.

DISCUSSION

Swyer-James-Macleod syndrome (SJMS) is a rare pulmonary disease which presents with unilateral hyperlucency of lungs. It results in decreased pulmonary vascularity and hyperinflation confined to one lobe or entire lung resulting from parenchymal destruction due to post infectious bronchiolitis obliterans. [1] Patients with Swyer-James syndrome often present with shortness of breath, decreased exercise tolerance, cough, hemoptysis and recurrent respiratory infections. [2] It is not uncommon that this disease may mimic bronchial asthma exacerbations or pulmonary embolism due to similar clinical symptoms. [3] Pulmonary hypertension complicating this syndrome is extremely rare and only few cases are reported in literature till date. Overlapping symptoms of breathlessness and cough, chest X-ray showing overwhelming pulmonary changes and pulmonary function test confirming obstructive lung pattern, leaves little room to suspect pulmonary hypertension in such a case. Clinical awareness of such an association is important as delay in diagnosis may lead to inappropriate therapy and progression of the disease resulting in increased morbidity and mortality of these patients. [4] We describe this rather unusual case of SJMS complicated with pulmonary hypertension which was managed with specific anti-PH therapy.

This patient was treated as bronchial asthma since childhood based on clinical symptoms with no symptomatic relief by many medical practitioners. The diagnosis of SJMS requires radiological evidence of unilateral or lobar hyperlucency of the lungs associated with decreased vascular markings, air trapping and variable degree of bronchiectasis. Chest CT is more sensitive to radiograph is detecting hyperlucent lung and associated findings. [5]

Initial diagnosis considered in our case was pulmonary embolism; however, CTPA ruled out its possibility and showed dilated MPA and RPA with atresia of left pulmonary artery. 2D echocardiography showed severe pulmonary hypertension with cor pulmonale. Although right heart catheterization (RHC) is the gold standard for determination of pulmonary pressures, but declined by family Pathogenesis of pulmonary hypertension is unclear in our patient, however possible mechanism might be vascular remodelling resulting from compensatory increase in blood flow in RPA and MPA due to hypoplastic pulmonary bed on left side. [6]

Although in our case clinical symptoms and lung function showed obstructive pattern suggesting asthma, chest radiograph showed increased lucency of left lung with decreased vascular markings and an absent hilar shadow suggesting Swyer-James syndrome which was confirmed on chest CT. Treatment of Swyer-James syndrome is mostly conservative which includes early control of respiratory infections and vaccination. Role of surgical intervention has been described in few cases with destroyed lung. [7]

Our patient had associated severe pulmonary hypertension with decreased exercise tolerance. We decided to start the patient on specific anti-PH therapy based on recommendation of recent guidelines. [8] Response to the therapy was modest and treatment was escalated as tolerated by the patient. Although uncommon, we referred our patient to undergo combined heart and lung transplant for definitive treatment in the face of persistent symptoms and imminent right heart failure. [9] Our case highlights the rare association of pulmonary hypertension with Swyer-James syndrome and its management approach. Although our patient has not undergone surgery yet, his outcome may provide more evidence on role of surgical intervention in Swyer-James syndrome.

REFERENCES

1. Lucaya J, Gartner S, García Peña P, Cobos N, Roca I, Liñan S. Spectrum of manifestations of Swyer-James-MacLeod syndrome. J Comput Assist Tomogr. 1998; 22(4):592.
2. Abba A, Al Mobeireek. Clinical spectrum of Swyer-James-Macleod syndrome in adults. Saudi Med. J. 2003; 24(2):195–198.
3. Walia M, Goyal V, Jain P. Swyer-James-Macleod syndrome in a 10-year-old boy misdiagnosed as asthma. Indian J Pediatr 2010; 77(6):709
4. J Henstenburg, N Patel, E Carnish. Dueling Lungs: The progression of Swyer-James Syndrome. The American Journal of Medicine. 2017; 7(9):485-486.
5. Moore A.D., Godwin J.D., Dietrich P.A., Verschakelen J.A., Henderson W.R., Jr. Swyer-James syndrome: CT findings in eight patients. Am. J. Roentgenol. 1992; 158:1211–1215.
6. M Yuce, N Bayram, O Ozer, V Davutoglu. An unusual cause of pulmonary hypertension: Swyer-James-Macleod syndrome. International Journal of Cardiology. 2016; 223:212-214.
7. Fregonese L, Girosi D, Battistini E, Fregonese B, Risso FM, Bava GL, Rossi GA. Clinical, physiologic, and roentgenographic changes after pneumonectomy in a boy with Macleod/Swyer-James syndrome and bronchiectasis. Pediatr Pulmonol. 2002; 34(5):412–416.
8. N. Galie, M. Humbert, J.L. Vachiery, et al., ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension, Eur Heart J. 2016; 37:167–219.
9. Hill C, Maxwell B, Boulate D, et al. Heart-lung vs. double-lung transplantation for idiopathic pulmonary arterial hypertension. *Clin Transplant.* 2015; 29:1067–1075.