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Medicine

ADULT DIAGNOSIS OF SWYER –JAMES-MACLEODS SYNDROME-A RARE CASE

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ABSTRACT Swyer-James-MacLeod Syndrome (SJMS) is a rare lung condition followed by bronchiolitis obliterans occurring in childhood . It is characterized by hypoplasia and/or agenesis of the pulmonary arteries resulting in hypoperfusion of the lung and unilateral hyperlucency on chest radiography . This entity is typically diagnosed in childhood with history of recurrent pulmonary infections but sometimes patients who have little or no symptoms may be missed till adulthood. Symptoms of decreased exercise tolerance, cough ,dyspnoea, and may be confused with bronchial asthma ,pulmonary embolism or bronchiactasis . Here we presented a 65year-old woman with SJMS discovered on the workup of acute onset breathlessness.

KEYWORDS : Swyer james Macleod,bronchiolitis,hyperlucency

INTRODUCTION:

In 1953, Swyer and James and, in 1954, Macleod, provided much more detail, described patients with unilateral hyperlucent lungs the latter providing more details. Swyer-James-Macleod syndrome (SJMS) also known as unilateral hyperlucent lung syndrome, is a rare long term complication of bronchiolitis obliterans usually after adenovirus infection occurring in infancy[1,2]. The child may be asymptomatic, but usually has recurrent pulmonary infections and develops bronchiectasis. Chronic productive cough , dyspnea and hemoptysis are some of the symptoms. A basic pathologic condition is bronchiolitis and obliteration of the small airways and alveolar destruction resulting in emphysematous pattern and dilated lung parenchyma[3]. As a result of inflammation the peripheral pulmonary vascularization is decreased. A chest radiograph shows a lobar or unilateral hyperlucent lung which may show normal or reduced volume of the affected lung. Pulmonary function testing (PFT) shows an air-flow and markedly decreased perfusion of the affected lung is revealed in the ventilation and perfusion scan [4].The bronchographic findings are remarkable with the major bronchi being normal, but the smaller branches appear club-like with small buds at the ends of the peripheral divisions. There is almost a complete absence of alveolar filling and a well-demarcated clear zone between the smaller bronchiectatic bronchi and the chest wall [5].This syndrome is typically diagnosed in childhood with recurrent respiratory infections, but those who have little or no symptoms or sequelae of bronchiectasis sometimes have minor may therefore not be diagnosed until adulthood[6].Here we present a 65year-old woman with SJMS discovered on the workup of acute onset breathlessness.

CASE REPORT

65 years old female with known case of rheumatoid arthritis , hemorrhoids came with complains of dyspnoea on exertion, loose motions, vomiting, loss of appetite, since 4 days. She had no history of fever or chest pain. She had a past history of recurrent pulmonary infections and productive cough. She was on steroid treatment for rheumatoid arthritis and no significant family history.

On general examination her pulse rate was 70/min, blood pressure was 110/70mmhg ,respiratory rate was 19 breaths per minute and oxygen saturation on room air was 88% .An elevated jugular venous pressure was present and mild bilateral pedal edema cyanosis ,clubbing or lymphadenopathy were absent.

On auscultation she had bilateral basilar crackles, air entry bilaterally decreased Her other systemic examination was normal.

She had a mild normochromic normocytic anemia with a leucocyte range of 7700/cumm and thrombocytosis with a count of 67800/mm^3. Her erythrocyte sedimentation rate was 104 and she had transaminitis with SGOT 83,SGPT57 and alkaline phosphatase 268,her total bilirubin and albumin levels were normal .Her C reactive protein levels were high 127.69,anti nuclear antibodies were positive but

rheumatoid factor was negative and uric acid levels and serum electrolytes ,creatinine and urea nitrogen was normal. Cardiac enzymes and the level of B-type natriuretic peptide were within normal range. An electrocardiogram showed sinus rhythm and normal axis. A chest X-ray (Figure1)showed hyperlucency in left lung field with paucity of markings as compared to the right showed a bony cage and soft tissues appear normal borderline cardiomegaly, CT ratio 13.2 : 24.3 cms. unfolding of aorta,linear atelectasis left lower zone,trachea was central and both costophrenic angles appeared normal,the hila, mediastinal and diaphragmatic outlines were normal . The transthoracic echocardiogram (TTE) with doppler showed normal sized cardiac chambers, normal Left ventricular systolic function,no regional wall motion abnormality(RWMA) ,Left ventricular ejection fraction 60 % and normal PA pressure .

Patient was treated with supplemental oxygen with a 24% face mask,bronchodilators ,hydroxychloroquine,steroids and probiotic capsules. A pulmonary test function (PFT) showed a obstructive-restrictive pattern.

A chest high-resolution computed tomography (HRCT) scan showed a diminished vascularity and hyperinflation pulmonary parenchyma and a mosaic perfusion defect on the left side, characteristically associated with SJMS.An extensive tree in bud opacities with discrete centrilobular alveolar opacities in basal segments of both lower lobes with a few similar opacities in medial segment of right middle lobe and in lingular segments suggestive of infective etiology(Figure 2).

Upper gastroscopy showed hiatus hernia and sigmoidoscopy was normal.

DISCUSSION

SJMS is characterized by unilateral hyperlucency of a part of or the entire lung considered to be a relatively uncommon and complex disease and was first described in 1953 by Swyer and James [7]. It is considered to be an acquired disease secondary to viral bronchiolitis and pneumonitis in childhood [8].The etiological organisms being Influenza A and adenovirus types 3, 7 and 21, Paramyxovirus morbillivirus, Bordetella pertussis, Mycobacterium tuberculosis, Mycoplasma pneumonia. Our patient had recurrent pulmonary infections in her childhood. Clinically, dyspnea on exertion, productive cough, sometimes with haemoptysis. Some patients, who have have minor symptoms or are asymptomatic and have no associated sequelae like bronchiectasis, therefore ,remain undiagnosed until they are adults.

In our case, SJMS diagnosis was reached during a dyspnoea workup done when the patent was 65 years of age. This diagnosis is based on the radiological pattern such as unilateral or lobar pulmonary hyperlucency with an air trapped lung during expiration . The affected lung parenchyma shows a variable degree of destruction and bronchiectasis could be associated. This could explain the obstructive-restrictive respiratory pattern typically seen on PFT and also reported in our patient. For this reason, the diagnosis of this syndrome is better established with HRCT on inspiration and expiration complemented with an angio-CT the latter could not be done in our patient. SJMS treatment includes the early control of lung infections as well as influenza and pneumococcal vaccinations [9,10]. No specific morbid-mortality studies with SJMS have been done.

CONCLUSION

SJMS is a post infective obliterative broncholitis with a variable clinical course and prognosis influenced by the presence of underlying bronchiectasis.The syndrome should be suspected in case of presumed pulmonary emphysema with an atypical distribution, pneumothorax or unilateral hyperlucency.

The most concerning long term effect of the syndrome is recurrent lung infection.Treatment of choice is symptomatic in conjugation with antibiotics. Surgery is only indicated in cases of unrelenting infection or the entire lung is affected. So the management comprises of chest physiotherapy inhaled corticosteroids and inhaled bronchodialator.

A hyperlucent lung field is an alarming radiographic finding. It should be considered as a differential diagnosis and diagnosed early,specially in patient not responding to conventional treatment to achieve better prognosis and also to prevent inappropriate treatment. Before drastic measures are initiated radiological images should be considered as guide both long and short term treatment options.

In summary, the present case emphasizes that a chest X-ray may underestimate the prevalence of the SJMS syndrome. The main reason for our reporting this case was related to the fact that few cases have been reported worldwide of adults presenting with SJMS.

LEGENDS

Figure 1 Hyperlucency In Left Lung Field With Paucity Of Markings As Compared To The Right

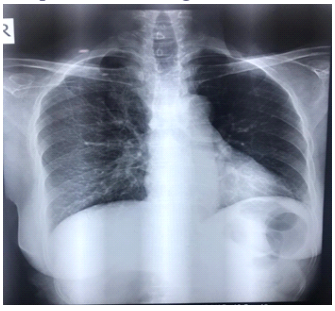
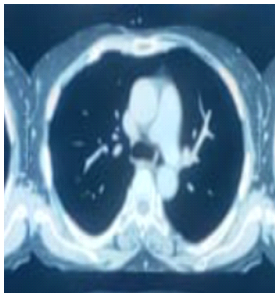


Figure 2 Chest High-resolution Computed Tomography (hrct) Diminished Vascularity And Hyperinflation Pulmonary Parenchyma An Extensive Tree In Bud Opacities With Discrete Centrilobular Alveolar Opacities In Basal Segments Of Both Lower Lobes.



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