CASE REPORT

SWYER JAMES (MACLEOD'S SYNDROME)

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Abstract: Macleod's Syndrome is a disorder of unilateral emphysema, bronchiecasis and loss of lung volume following bronchiolitis occurring or patchy bronchitis in infancy or childhood. We describe a case who presented with polycytemia and corpulmonale; thisunusual presentation has not been reported so far.

INTRODUCTION

This is a ratre disorder characterized by unilateral Emphysema of a lung or resulting from localized bronchiolitis or bronchitis following viral infections or tuberculosis during infancy or child hood. Radiologically there is unilateral hyper translucency with diminished pulmonary vascular markings and normal or reduced lung vlume and bronchiectasis [1-4]. The disorder was initially described by Swyer and James in 1953 2 and by Macleod a year 4 and subsequently few more cases were reported. We describe the syndrome in a 16 year old male who presented with polycythemia and cor pulmonale possibly representing the first case of such nature in literature.

CASE REPORT

A sixteen year old male was admitted with history of productive cough and dyspnea since isx months of age with frequent hospitalizations. On examination he was grossly cyanosed, with pectus carinatum and features of corpulmonale with right heart failure. On investigations he had hemoglobin of 18.5 gm/dl and hematocrit of 66%. Chest radiograph revealed evidence of bilateral bronchiectasis, emphysema and reduced lung volume on rith side and cadiac enlargement of right ventricle (figure1). Electrocardiogram revealed right axis deviation p-pulmonale, and R in V1 and V2 High resolution computed tomography (HRCT) showd bilateral bronchiectases, and hyper lucent lung fields with emphysema on right side (Figure2) Echocardiography demonstrated pulmonary arterial hypertension, tricuspid regurgitation and other findings of cor pulmonale. On pulmonary function testing he had mild obstruction. The serum levels of alpha one antitrypsin were normal and all other investigations including serum electrophoresis and immunoglobulin levels were normal. Arterial blood gas analysis revealed desaturation (68% and respiratory acidosis. The patient was managed with oxygen, diuretics, phlebotomies and other supportive measures. He improved symptomatically and clinically and is under regular follow up.

DISCUSSION

On account of rarity, there is paucity of literature regarding the disorder. It seems likely that this disorder starts between six months to five years of age after viral broncholitis or tuberculosis, and is characterized by cough, dyspnea, hemoptysis and clinical manifestations of bronchiectases [1.3.4]. The affected lung is usually subnormal in size because of the subsequent underdevelopment of lung after bronchiolitis. If the disorder occurs early in life, the lung fails to grow normally and remains smaller than the opposite lung but if it occurs later in childhood, the lungs may be of normal size [1.4]. On histopathological examination there is pancinar emphysema, with fever alveoli than normal. The pulmonary artery is often hypo plastic on angiography possibly due to local hypoxemia. There is often hypertrophy of the pulmonary artery walls and a decreased number

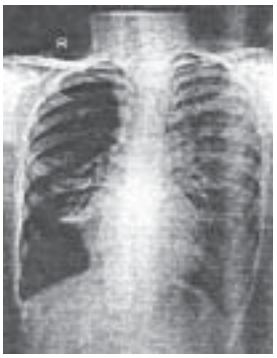


Figure-1*:Postero anterior view of chest readiography showing bilateral bronchiectatsis, emphysema on right side and features of cor pulmonale.*



Figure-2 :*HRCT showing bilateral bronchiectasis and emphysema on the right side*

of branches. On pulmonary function testing there is often evidence of airway obstruction and an increase in the residual volume. Bronghography demonstrates diffuse bronchiectasis with absence of filling of the terminal bronchioles. Radionuclide one scanning shows decreased ventilation and persuasion in the affected area. HRCT is considered as investigation of choice for identifying the characteristic changes [2.5]. The disorder needs to be differentiated from compensatory emphysema, where the collapsed opposite lobe can by visualized and the vessels in emphysematous lobe are fanned out. The occurrence of cor pulmonale as in our case is most likely due to hypoxic vasoconstriction following emphysema and the diminished vascular cross sectional area from bronchiectasis [6] In future further studies are needed regarding etiopathogenesis, treatment modalities, and prognosis of the disorder.

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