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A Case Series of Corpulmonale Secondary to Sle as A Rare Cause

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ABSTRACT *Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that has protean manifestations and follows a relapsing and remitting course. More than 90% of cases of SLE occur in women. Incidence of corpulmonale is very rare in a case of SLE, although documented in other autoimmune disorders secondary to pulmonary arterial hypertension. Here we report 5 cases of systemic lupus erythematosus (SLE) who developed clinical manifestation of pulmonary hypertension and corpulmonale. These patients presented with different manifestations like dyspnea, peripheral edema, oral ulcerations, photosensitivity . Clinically significant pulmonary hypertension is a rare complication of SLE and increased pulmonary vascular resistance may not be entirely fixed.*

Introduction:

SLE is commonly associated with both pulmonary and cardiac pathology. Although approximately one third of patients develop pleural effusion involvement of lung parenchyma is somewhat less common , with radiological findings varying from increased bronchial markings to diffuse interstitial infiltrates(1).Cardiac involvement may consist of pericarditis, myocarditis and non infectious endocarditis(2).Pulmonary hypertension and cor pulmonale have been seen in collagen vascular diseases, particularly scleroderma essentially in association with pulmonary infiltrates(3).There have been fewer cases reported of such involvement in SLE. We report about a patient with SLE who developed severe pulmonary hypertension, despite the absence of pulmonary infiltrates, during a period when her disease was otherwise in clinical remission.

Case series:

We report 5 different cases of SLE with features of pulmonary arterial hypertension and cor pulmonale who presented to us differently. The first case being a 25 year old female presented with history of erythematous rash over face, easy fatigability onset 3 years back. Later after 1 year patient developed exertional class II NYHA breathlessness,bilateral lower limb swellings. On examination she was thin with malar rash over face, hyperpigmented lesions on extensor aspect of forearms. Cardiac examination revealed a normal apical impulse, loud second heart sound with 2/6 ejection systolic murmur at pulmonary area. ANA and anti-dsDNA was positive and on 2D Echo showed dilated right atrium and ventricles, moderate tricuspid regurgitation with TR jet 45mm of Hg and mild mitral regurgitation, pulmonary artery diameter was 3.4cm.

Second patient a 39 year old female presented with complaints of dyspnea of class III NYHA and swelling of feet and abdominal distension. On examination loud S2 and ejection systolic murmur was present in Pulmonary area. Electrocardiogram revealed right dominance with right axis deviation.ANA and anti-dsDNA was positive and on 2D Echo showed dilated right atrium and ventricles, moderate tricuspid regurgitation with TR jet 55mm of Hg and, pulmonary artery diameter was 3.9cm.

Third patient a 29 year female, who is a diagnosed case of SLE on follow up presented with similar complaints of dyspnea and occasional palpitations. On auscultating loud S2 present with afaint murmur. 2D Echo showed dilated right atrium and ventricles, moderate tricuspid regurgitation with TR jet 40 mm of Hg, pulmonary artery diameter was 3.3cm.

Fourth patient a 36 year old female, again a known case of SLE on follow up presented with complaints of swelling of feet and abdominal distension with easy fatiguability and exertional dyspnea. 2D Echo showed dilated right atrium and ventricles, moderate tricuspid regurgitation with TR jet 50 mm of Hg and , pulmonary artery diameter was 4cm.

Last patient in the series who is a 27 year old female presented with complaints of photosensitivity and dyspnea and pedal edema. On examination oral ulcerations, alopecia and rash present. Cardiovascular examination showed a raised JVP with loud S2. ANA and anti-dsDNA was positive and on 2D Echo showed dilated right atrium and ventricles, moderate tricuspid regurgitation with TR jet 60mm of Hg and mild mitral regurgitation, pulmonary artery diameter was 3.9cm.

Thus from the avove evidences all the patients were concluded as having systemic lupus erythematosus with pulmonary hypertension with corpulmonale. They were treated with hydroxychloroquine 100mg twice daily for SLE, Tab.Lasix 40mg od, Tab.Sildenafil 20mg bd for pulmonary hypertension and showed symptomatic improvement and were under follow up.

Discussion:

SLE is a chronic autoimmune disease that can affect almost any organ system; thus, its presentation and course

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are highly variable, ranging from indolent to fulminant. The classic presentation of a triad of fever, joint pain, and rash in a woman of childbearing age should prompt investigation into the diagnosis of SLE. The diagnosis of SLE is based on a combination of clinical findings and laboratory evidence. Familiarity with the diagnostic criteria helps clinicians to recognize SLE and to subclassify this complex disease based on the pattern of target-organ manifestations. The presence of 4 of the 11 American College of Rheumatology (ACR) criteria yields a sensitivity of 85% and a specificity of 95% for SLE

Although the specific cause of SLE is unknown, multiple factors are associated with the development of the disease, including genetic, epigenetic, ethnic, immunoregulatory, hormonal, and environmental factors. Many immune disturbances, both innate and acquired, occur in SLE. Many clinical manifestations of SLE are mediated by circulating immune complexes that form with antigens in various tissues or the direct effects of antibodies to cell surface components. Immune complexes form in the microvasculature, leading to complement activation and inflammation. Moreover, antibody-antigen complexes deposit on the basement membranes of skin and kidneys

Pulmonary hypertension is thought to be a rare complication of SLE , but may occur more commonly in other collagen vascular diseases , notably in systemic sclerosis and mixed connective tissue diseases(4).It was postulated that pulmonary vascular degeneration, thrombosis and resultant pulmonary infarction might be the basis for episodes of unexplained breathlessness without evidence of pulmonary parenchymal infiltrates that occur in some patients with SLE as in our case.

Severe pulmonary hypertension in association with SLE may present at a time when other manifestation of SLE are quiescent(5).This suggest that an initial insult to the pulmonary vasculature might take several years to produce overt pulmonary hypertension. These was true not only of our patient with SLE but also of those reported by Charoerpar et al (5) and yeo and sinnah and in the patient with mixed connective disorder reported by jones et al (4).

Our patients also illustrates the point that patient with SLE may present with pulmonary hypertension without any evidence of parenchymal lung disease by chest X ray.

A retrospective study in Singapore identified 3% of 786 lupus pts with primary and secondary pulmonary hypertension measured by TTEC. A prospective clinical and echo cardiography study performed on 84 disease patients with lupus showed Pulmonary arterial hypertension in 11% of cases.

Conclusion:
SLE a disease of autoimmunity presents variedly and is associated with a wide range of complications with multi system involvement. Pulmonary hypertension seen in many autoimmune diseases is a very rare entity in SLE.

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