RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 10 | October 2014 | ISSN - 2249-555X

SLE - A Rare Cause of Cor Pulmonal – a Case Report

KEYWORDS: Pulmonary hypertension, Systemic lupus erythematosus

Dr. K.M.Iqbal Hussain
M.D., I/C Professor of Medicine,Government General Hospital, Kurnool.

Dr. Lakshmi Bai
M.D., Associate Professor of Medicine, Government General Hospital, Kurnool.

Dr. Pullaiah
, Assistant professor in General Medicine, Government General Hospital, kurnool.

Dr.Teliki Vinil
Resident in general Medicine, Government General hospital ,Kurnool. Phone- 9052551101

ABSTRACT *A Case is reported of a 25 year old woman with systemic lupus erythematosus (SLE) who developed clinical manifestation of pulmonary hypertension at when other manifestation of SLE were quiescent.Clinically significant pulmonary hypertension is a rare complication of SLE and increased pulmonary vascular resistance may anot be entirely fixed .*

Introduction:
SLE is commonly associated with both pulmonary and cardiac pathology. Although approximatly 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 report:
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.Patient had no history of smoking, oral contraceptive pills usage. Home maker by occupation.

On examination patient was thin with malar rash over face, hyperpigmented lesions on extensor aspect of forearms. Her pulse rate was 88 beats/min bloodpressure 110/80mm Hg, temperature - 98.4^0 F, respiratory rate - 28/min. On auscultation lungs were clear. Cardiac examination revealed a normal apical impulse, loud second heart sound with 2/6 ejection systolic murmur at pulmonary area. Mild splenomegaly on palpation.There was no joint deformities

Routine laboratory finding included an antinuclear antibody (ANA) which was detected in serum at quantity of 88.77 Units and anti-dsDNA antibody was 272.25 IU/ml. Serum complement levels was normal. Antiphospholipid antibodies were negative. Chest X ray PA view showed cardiomegaly with right ventricular prominence and enlargement of proximal pulmonary arteries.

Electrocardiogram showed poor 'R' wave progression. Echocardiogram 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.High resolution computerized tomography of chest showed mildly dilated pulmonary trunk, right and left pulmonary arteries.Thus from the above evidences patient concluded as having systemic lupus erythematosus with pulmonary hypertension with corpulmonale.Patient was treated with hydroxychloroquine 100mg twice daily for SLE, Tab.Lasix 40mg od, Tab.Sildenafil 20mg bd for pulmonary hypertension.Patient showed symptomatic improvement.

Fig.1.malar rash

Volume : 4 | Issue : 10 | October 2014 | ISSN - 2249-555X

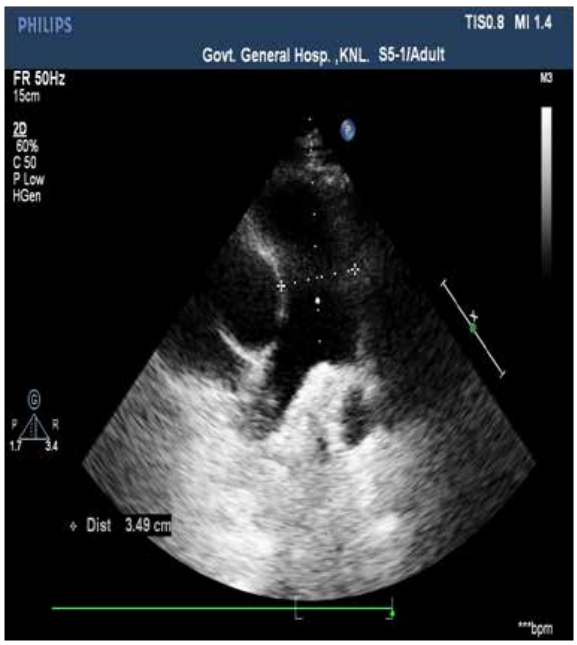

fig.2.2DECHOshowing dilated pulmonary artery

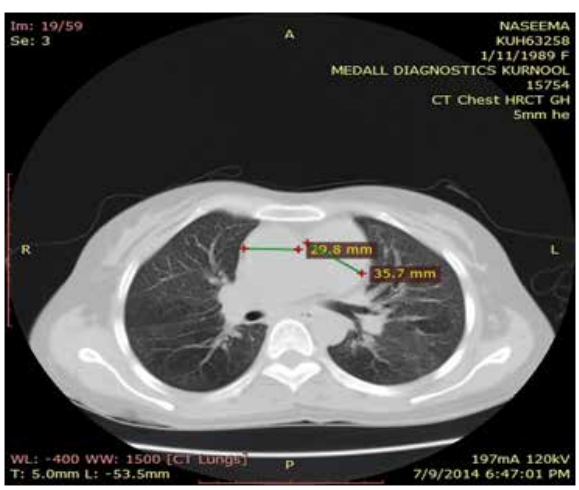

Fig3.HRCT chest showing dilated pulmonary artery

Discussion:

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 patient 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:

connective tissue disorders rarely causes pulmonary arterial hypertension and corpulmonale. Secondary causes should be ruled out before labeling as primary pulmonary hypertension.High index of suspicion and complete investigative work up is necessary to make a diagnosis and choose modality of treatment.

REFERENCE

• 1) Charoenpan P.sukumaichantra y, AyuthyaWIN:- pulmonary hypertension in systemic lupus erythematosus. A case report J.med assoc thai 60, | • 2) hejtmanciks M, Wright J, quint R , jenninF The cardiovascular manifestation of SLE , Am heart J 68,. | • 3) Cummings P:- Primary pulmonary hypertension and SLE , N.England J.med 288,1078. | • 4) Ester D Christian CL:- The Natural history of SLE by prospective analysis. | • 5) Faymei A:- Pulmonary vascular disease in SLE : AJ clin.pathology.. | • 6) RuskinJN,Hunter AM: PPH treated with pral phentolamine. | • 7) Levine BW,gang :- case reording of the massachusetts general hospital. | • 8) Jones H, osterholm R, Wilson R, Martin F, Commer T .Bachmayer T, Fatal pulmonary hypertension & resolving immune complex glomerulonephritis in MCTD