World Journal of Pharmaceutical and Life Sciences
WJPLS
www.wjpls.org

PULMONARY THROMBOEMBOLISM AS A FIRST PRESENTATION OF SYSTEMIC LUPUS ERYTHMATOUS.

[1]Lalit Prashant Meena[1*], Jaya Chakravarty[2], Saurabh Chopra[3] and Madhukar Rai[4]

[1]Assistant Professor, Department of Medicine, Institute of Medical Sciences, BHU, India.

[2]Associate Professor, Department of Medicine, Institute of Medical Sciences, BHU, India.

[3]Jr, Department of Medicine, Institute of Medical Sciences, BHU, India.

[4]Professor, Department of Medicine, Institute of Medical Sciences, BHU, India.

Article Received on 26/09/2015 Article Revised on 17/10/2015 Article Accepted on 08/11/2015

***Correspondence for Author**
Dr. Lalit Prashant Meena
Assistant Professor, Department of Medicine, Institute of Medical Sciences, BHU, India

ABSTRACT

Background: Systemic lupus erythematous (SLE) can involve any organ in body, but pulmonary vascular system involvement is usually in the latter course of the disease. **Case Report:** Here we are submitting a case of SLE who was presented with massive pulmonary thrombosis as a presenting symptom. A 25 year old unmarried female patient presented to us with complaints of sudden onset breathlessness and retrosternal chest pain. On the basis of examination and investigations SLE with Pulmonary thromboembolism was diagnosed. She was treated with Immunosuppressive and anticoagulant therapy and showed a dramatic good response. **Conclusion:** Patient comes with sign and symptoms of Pulmonary thromboembolism as a first manifestation, should also be evaluated for SLE and Anti phospholipid antibody.

KEYWORDS: Systemic lupus erythematous (SLE), pulmonary vascular system.

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease that primarily affects female of childbearing age with a 10:1 female to male ratio.[1] Any organ in body can be affected by SLE; but pulmonary involvement is usually in the latter course of the disease.[2,3,4] In a study, 25% of patients with SLE had clinical and/or radiographic evidence of pulmonary involvement.[5] Here we present a case of SLE who came with the symptoms of massive pulmonary thrombosis as a presenting symptom.

CASE REPORT

A 25 year old unmarried female patient presented to us with the complaints of sudden onset breathlessness and retrosternal chest pain for 5 days. Patient was diagnosed as pulmonary koch's 6 months back for similar complaints and has been on Anti tubercular treatment since then. On retrospective history she also had the complains of multiple joint pain and skin rashes one and half year back. There was no history of fever, cough, weight loss or loss of appetite, hemoptysis, orthopnea, paroxysmal nocturnal dyspnea, pain in legs or calf region and similar illness in the family. Her menstrual cycles were regular and there was no history of foetal loss. On examination mild to moderate pallor, pitting edema and Left axillary lymphadenopathy were present. Jugular venous pressure was raised. Respiratory rate, Pulse Rate and blood pressure were 24/min, 96/min,and 112/74mmhg respectively. On Chest examination bilateral fine crepitations were present with diminished air entry in left infra axillary region .On cardiovascular examination P2 was loud and a Pansystolic murmur was audible at tricuspid area. Abdomen was soft, nontender with mild tender hepatomegaly .Her blood investigations showed Hb 6.3 gm/dl, TLC 5400/mm^3, PLT 4.22 lacs/ mm^3, Cr/Ur 1/79 mg/dl, Na /K 139/4.2 mmol/l, SGOT/PT 18/47 U/L, TB/DB 0.4/0.1mg/dl, ALP 103 U/L, TP/ALB 7.0/2.8 gm/dl, Fe/TIBC 11/187µmol/l, LDH 749 U/L. ABG analysis showed hypoxemia with pO2 62.9 mmHg, pCO2 36.9 mmHg, SO2 92.2 mmHg, HCO3 19.9 mEq/l. Her Sputum for AFB was Negative. Urine routine microscopy showed trace albuminuria, 4-6 pus cells per high power field and 24 hour Urine Protein estimation was 273.6 mg /24 hours. Her coagulation profile showed Prothrombin time 15.1 with INR 1.33, APTT 28.3 with control 30.0 and raised D-Dimer (1200 ng/ml). Autoimmune profile showed RA Factor 70.0 IU /ml (N <20 IU /ml), CRP titre 4.45 mg/ dl (N <0.6 mg /dl), Anti CCP 2 Ab -9.0 u/ml (N <25.0 u / ml), ANA -Positive (titre - 1:1280), Anti Ds DNA Ab 153.2 IU /ml (N35.0 IU /ml). Anti Cardiolipin antibodies:- IgG 7.79 GPL U/ml (negative), IgM 14.2 MPL U/ml (weakly positive), IgA 6.28 APL U /ml (negative). Antiphospholipid antibodies - IgG- 12.3 U/ml (weakly positive), IgM-18.2 U/ml (weakly positive), Beta 2 Glycoprotein (EIA) was Positive. IgG - 4.96 (n<20.0), IgM -93.07 (n<20.0) , IgA 41.80 (n<20.0) .Her ECG showed Sinus Tachycardia, T wave inversion in leads v1 to v4 and 2D Echocardiography revealed severe TR,severe PAH with mild MR,normal LA/LV , Dilated RA/RV ,normal LV systolic and diastolic function, mildly hypokinetic Right Ventricle, LVEF 63% ,no clot / thrombus. PASP 84 mmHg RVSP 74mmHg. Colour Doppler of b/l lower limbs revealed no evidence of any thrombus. X-Ray chest showed the features of severe pulmonary artery (Image 1). CT thoracic angiography showed thromboembolism in segmental branch of right pulmonary

artery (posterior basal segment) with associated air space opacification noted in involved segment suggestive of infarct. Thrombus also evident in right middle pulmonary artery branch & segmental branch of left main pulmonary artery (post. Basal segment of left upper lobe) with Moderate to severe hepatic steatosis and few hypodense foci in upper pole suggestive of splenic infarct (Image 2). Patient was diagnosed as SLE with Pulmonary thromboembolism with Pulmonary artery hypertension. She was treated with systemic steroid, Cyclophosphamide and with anticoagulants. On follow up patient showed a dramatic improvement in sign and symptoms and in chest x-ray findings (Image 3).

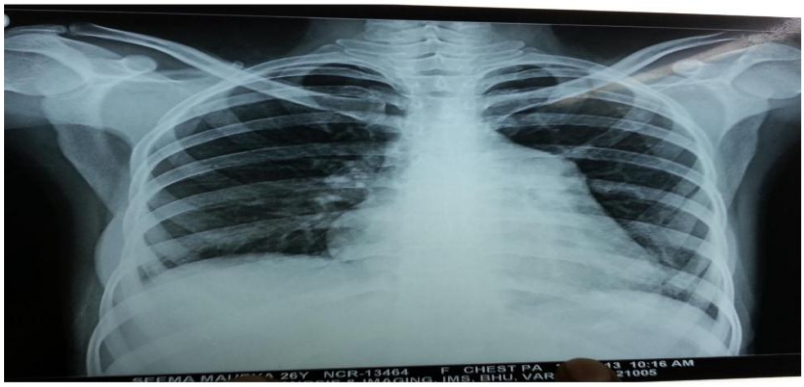

Image. 1: X-Ray Chest at the time of presentation showing the prominence of Right Pulmonary artery (before treatment)

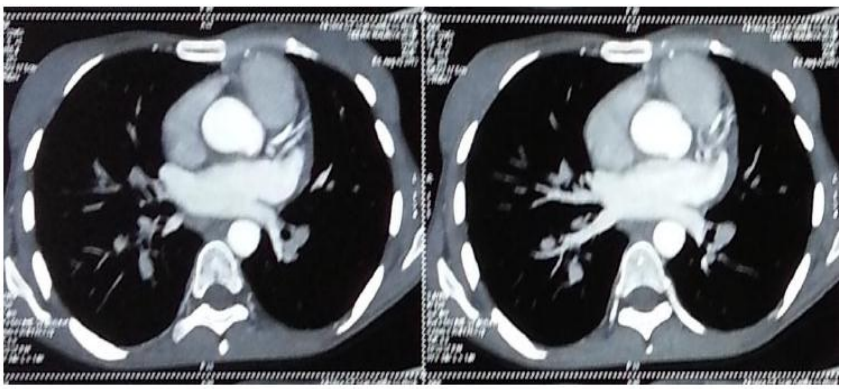

Image.2

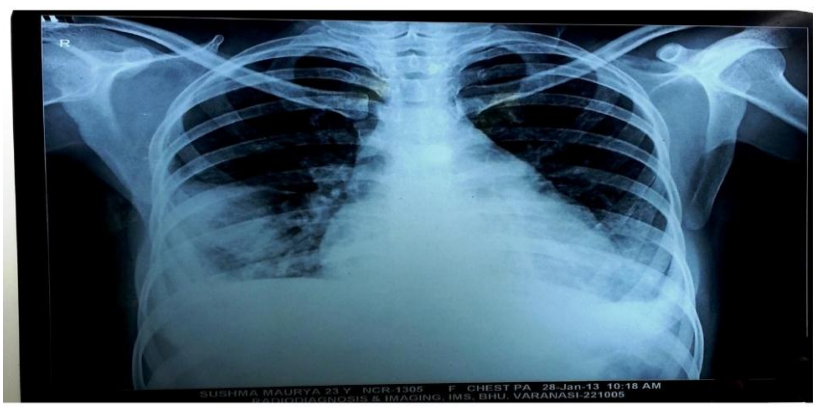

Image.3: X-RAY Chest after treatment

CT THORACIC ANGIOGRAPHY

Thromboembolism in segmmental branch of right pulmonary artery (posterior basal segment) with associated air space opacification noted in involved segment s/o ? Infarct.

Thrombus also evident in right middle pulmonary artery branch & segmental branch of left main pulmonary artery (post. Basal segmt. Of left upper lobe Moderate to severe hepatic steatosis Few hypodense foci in upper pole s/o splenic infarct.

DISCUSSION

Patients with Systemic lupus erythmatous are at increased risk of VTE (venous thromboembolism) with a prevalence rate of 9%.[6] It is mostly related to disease activity. Patients with antiphospholipid antibodies have more increased risk reaching up to 35% to 42%. Venous thromboembolism can occur either acutely (deep vein thrombosis or acute pulmonary embolism) or chronically resulting in chronic thromboembolic pulmonary hypertension. Clinical presentations of Systemic lupus erythmatous associated Pulmonary artery hypertension (PAH) is similar to idiopathic pulmonary arterial pulmonary hypertension (IPAH). Symptoms include dyspnea, fatigue, chest pain and lower limb swelling. Physical examination includes jugular venous distension with a large V wave, loud pulmonic component with wide splitting of the second heart sound, murmur of tricuspid regurgitation and/or pulmonic insufficiency, and lower limb edema. Physical findings may be minimal in mild Pulmonary hypertension. The prognosis of SLE associated PAH is worse than IPAH, with a 5-year survival of only 17% compared to 68% in patients with IPAH.[7] Long term anticoagulation with warfarin and a target INR of 2.0 to 3.0 is highly recommended .High intensity warfarin (target INR 3.0-4.0) was not found to be superior to moderate intensity warfarin (target INR 2.0-3.0). Moderate intensity warfarin had lower rate of major bleeding.[8] Any organ in body can be affected by SLE; but pulmonary involvement is usually in the latter course of the disease.[2,3,5] But in our case the patient presented primarily with symptoms of pulmonary thromboembolism without DVT at other vascular system of body which is a very rare manifestation of SLE.

CONCLUSION

Although in literature it is described that Pulmonary artery hypertension due to thromboembolism in SLE developed in late stage, but like our case if a female patient comes with sign and symptoms of Pulmonary thromboembolism as a first manifestation ,should also be evaluated for SLE and Anti phospholipid antibody.

REFERENCES

1. Siegel, M., & Lee, S. L. The epidemiology of systemic lupus erythematosus. Semin Arthritis Rheum, 1973; 3: 1-54.
2. Haupt, H. M., Moore, G. W., & Hutchins, G. M. The lung in systemic lupus erythematosus. Analysis of the pathologic changes in 120 patients. Am J Med., 1981; 71: 791-798.
3. Orens, J. B., Marti nez, F. J., & Lynch, J. P.,3rd. Pleuropulmonary manifestations of systemic lupus erythematosus. Rheum Dis Clin North Am., 1994; 20: 159-193.
4. Quadrelli, S. A., Alvarez, C., Arce, S. C., Paz, L., Sarano, J., Sobrino, E. M., & Manni, J. Pulmonary involvement of systemic lupus erythematosus: analysis of 90 necropsies. Lupus, 2009; 18: 1053-1060.
5. Pego-Reigosa, J. M., Medeiros, D. A., & Isenberg, D. A. Respiratory manifestations of systemic lupus erythematosus: old and new concepts. Best Pract Res Clin Rheumatol, 2009; 23: 469-480.
6. Gladman, D. D., & Urowitz, M. B. Venous syndromes and pulmonary embolism in systemic lupus erythematosus. Ann Rheum Dis., 1980; 39: 340-343.
7. Chung, S. M., Lee, C. K., Lee, E. Y., Yoo, B., Lee, S. D., & Moon, H. B. Clinical aspects of pulmonary hypertension in patients with systemic lupus erythematosus and in patients with idiopathic pulmonary arterial hypertension. Clin Rheumatol., 2006; 25: 866-872.
8. Crowther, M. A., Ginsberg, J. S., Julian, J., Denburg, J., Hirsh, J., Douketis, J. et al. A comparison of two intensities of warfarin for the prevention of recurrent thrombosis in patients with the antiphospholipid antibody syndrome. N Engl JMed., 2003; 349: 1133-1138.