Abstract

Background

Mixed connective tissue disorder (MCTD) is a rare multi-systemic collagen vascular disorder characterized by the overlapping features of systemic lupus erythematosis, systemic sclerosis, polymyositis/dermatomyositis, and rheumatoid arthritis with the unique and invariable presence of the Anti-U1RNP antibodies. Although the disease follows an indolent course with regard to renal and neurological morbidity, it confers significant mortality due to pulmonary hypertension and interstitial lung disease. Thromboembolism due to antiphospholipid syndrome is much less common in MCTD and much less the chronic pulmonary thromboembolism contributing to pulmonary hypertension.

Case

This is a case of a 43 year old female with a history of palindromic rheumatism presenting and severe fatigue with progressive dyspnea over six months with features of pulmonary hypertension and recent onset small and large joint synovitis, hand oedema ,raynauds phenomenon with difficulty in walking due to myopathic weakness.

Conclusion-

Mixed connective tissue disorder can present with chronic pulmonary thromboembolism contributing to pulmonary hypertension.