

## Pleuropericarditis an unusual preliminary manifestation of Mixed Connective Tissue Disorder

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### Abstract

*Though cardiac involvement has been observed in mixed connective tissue disorder(MCTD). Pleuropericarditis with or without an effusion is rarely seen as a preliminary manifestation of MCTD. We report a 40 year old patient with pleuropericarditis who was diagnosed with MCTD after having a high index of suspicion with the aid of diagnostic tools. This report highlights the importance of screening for connective tissue disorders in patients with pleuropericarditis.*

**Keywords:** *Pleuropericarditis, Raynaud's phenomenon, Anti RNP antibodies.*

### Introduction

The most common manifestations of MCTD are arthralgias, Raynauds phenomenon, swollen joints (sausage like appearance) esophageal dysfunction, pulmonary hypertension and muscle weakness. It was first described in 1972 by Sharp et al as a syndrome with mixed features of various connective tissue disorders such as systemic lupus erythematosus (SLE), systemic sclerosis(SSc), rheumatoid arthritis(RA), dermatomyositis (DM) and polymyositis (PM) along with moderate to high titre of anti-U1 small nuclear (sn) anti-ribonucleoprotein (anti-RNP) antibodies, high speckled fluorescent antinuclear antibodies. The most common requirement for diagnosis is the serological criteria and at least three clinical criteria. Extensive organ involvement may occur and has been associated with bad prognosis. Pleuropericarditis usually responds to NSAIDS and or corticosteroids depending on its severity[1-2].

### Case report:

A 40 year old male patient not a known case of any major illness presented with the complaints of heaviness of

chest, breathlessness and bodyache symptoms gradually progressed not relieved by rest since three days. He was a chronic tobacco chewer and occasional alcoholic and smoker. On examination he was afebrile normotensive and systemically nothing abnormal was detected except decreased air entry in the bases bilaterally.

On investigating, his electrocardiogram showed concave upwards ST elevations in all leads, his hemoglobin was 11.8 and total leukocyte count was within normal limits. His blood sugar levels, lipid profile, renal function tests including serum electrolytes and liver function tests were normal. Troponin T was negative. His chest Xray revealed that the left cardiophrenic angle was blunted and ultrasonography of abdomen was normal while chest sonography revealed bilateral mild pleural effusion with underlying consolidation, antibiotics were added.

Patient became increasingly breathless his arterial blood gases showed respiratory acidosis and his chest pain increased, especially in lying down position and relieved on sitting up and he complained of abdominal pain. On auscultation he had bilateral crepitations, pericardial rub with reduced air entry in the bases bilaterally. His repeat chest Xray showed a flask shaped enlargement of the heart shadow and sputum AFB and gram stain were normal. His D Dimer and amylase levels were normal. His 2D echo showed minimal pericardial effusion with trivial tricuspid regurgitation with no pulmonary hypertension and ejection fraction showing 60 percent and no RWMA(Regional wall motion abnormality). His Erythrocyte sedimentation rate was raised 86. ANA was positive more than 2.5. His RNP was positive using the

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U1-RNP-specific antigen enzyme-linked immunosorbent assay test. Patient was started on prednisolone 1mg/kg. On further enquiry history revealed a Raynauds phenomenon like symptoms in the past. Serial Electrocardiograms showed persistent concave upwards ST elevations in all leads. CT Thorax confirmed scanty pleural effusion with subsegmental left anterior basal underlying consolidation of the lung, pericardial effusion. Viral markers testing was advised to the patient but we were unable to obtain this test.



**Fig 1:** CT Thorax confirmed scanty pleural effusion with subsegmental left anterior basal underlying consolidation of the lung, pericardial effusion.

## Discussion

Mixed connective tissue disorder is characterized by features seen in Rheumatoid disease, Scleroderma, SLE and dermatomyositis. Ninety percent female preponderance is seen with a ratio of 10:1 and second and third decades show the highest incidences. Though the presence of pleuropericardial disease in connective tissue disorders is common but a preliminary presentation of pleuropericarditis with or without effusion is a rarity. Constrictive pleuropericarditis and cardiac tamponade is rarely seen[3].

Pleuropericardial disorders occurring in connective tissue diseases are not uncommon and may present as acute or chronic pericarditis with or without an effusion but presenting as the initial manifestation is a rarity. In most cases the diagnosis of pericarditis is found only during autopsy. The pericardial involvement is seen in 59% cases in scleroderma, 44% in SLE, 30% mixed connective tissue disorders, 24% rheumatoid arthritis and in 115 cases of polymyositis and dermatomyositis according to averages taken in various studies. Cardiac tamponade or constrictive pleuropericarditis are rarely seen[4].

Oetgen et al noted a 38% incidence of cardiac abnormality out of which 25% had pericarditis[5]. While Singesen et al noted a 43% incidence of pericarditis in mixed connective tissue disorder(MCTD)[6]. The treatment of pericarditis included non steroidal anti-inflammatory drugs (NSAIDS), corticosteroids, pericardial tapping and in some cases resection of pericardium in constrictive pericarditis.

MCTD is a distinct clinical entity with a wide gamut of clinical manifestations. The high mortality and worst prognosis occurs with the occurrence of pulmonary disease especially pulmonary arterial hypertension. Cardiac involvement varying from 11% to 85% sees pericarditis in 10% to 29% cases, in 26% cases myocarditis, mitral valve prolapsed, conduction disturbances(heart block), diastolic filling failure are seen. The most common modality for diagnosis is the serological and three clinical criteria. Treatment consensus is not specific but should be tailor made to suit each individual case and aggressive treatment for medical emergencies should be anticipated and employed accordingly[7].

Interstitial lung disease is the most common finding in MCTD(21% to 66%) and most frequently seen is the NSIP(Non specific interstitial pneumonia). Ground glass opacities in the peripheral lower lung fields. Pleural thickening and effusion is seen only in 10 % cases. Other pulmonary features of MCTD are pneumonia, pulmonary vasculitis, thromboembolism, alveolar hemorrhage and pulmonary hypertension which is the most important cause of death. Corticosteroids remains the main line of treatment with a good response seen in MCTD cases. This conventional treatment fails or patient develops intolerance. Colchicine and sometimes immunosuppressives like azathioprine or cyclophosphamide may be added for recurrent pericarditis[8].

## Conclusion

The pericardial disorders occurring in various connective tissue disorders is rarely fatal but one needs to be on a watch for symptoms and signs indicating deterioration and institute prompt treatment of cardiac tamponade. Pericarditis is the most common manifestation, pericardial effusion is not so common and cardiac tamponade is rarely seen only in well established cases of MCTD. The wide use of steroids and NSAIDS decreases the incidence of tamponade despite the high incidence of pericarditis. MCTD should be included in

the treatable causes of pleural effusions. Thus the diagnosis of MCTD should be kept in mind by treating physicians in cases of pleuropericardial effusions when all other causes are ruled out.

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