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## Case report on patient with Interstitial Lung Disease, Pulmonary Hypertension and POEMS Syndrome.

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### ABSTRACT

Interstitial Lung Disease is a group of lung diseases with two-thirds of unknown etiology and remaining one-third either associated with connective tissue disorder or caused by various environmental or occupational exposures. POEMS Syndrome is a multi-system disorder associated with polyneuropathy, organomegaly, endocrinopathy, a monoclonal protein (M Protein) and skin changes.[1] The author describes a patient with Interstitial Lung Disease , Pulmonary Hypertension and POEMS Syndrome who presented with hepato-splenomegaly , increased blood sugar levels , Ascites , Pleural Effusion and skin hypopigmentation.

**Keywords:**lung, pulmonary hypertension, POEMS syndrome.

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**CASE SUMMARY:**

A patient 49 years male, a cook by profession came with complaints of non-productive cough since 2 years (on and off), breathlessness since 2 weeks, Grade IV MMRC, Left sided chest pain-non radiating and increased on coughing and in deep inspiration, abdominal distension, swelling of both lower limbs and facial puffiness.

On examination, patient had generalised anasarca, hypopigmentation of the skin on both legs and hands, clubbing, white nails and bilateral pitting pedal odema. Patient had a saturation of SP02-79.2% at room air.

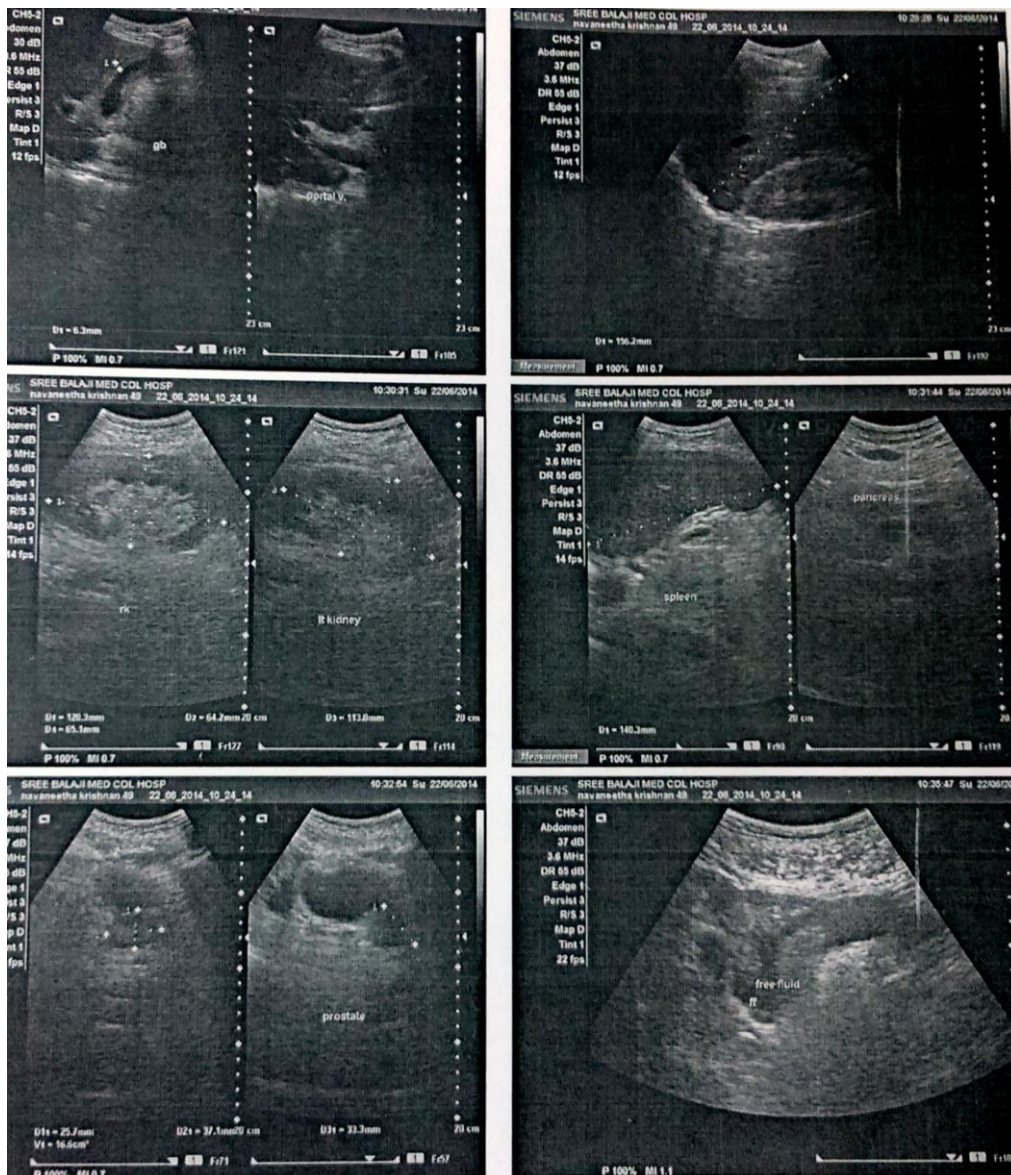
Fluid Thrill and shifting dullness were positive on Abdominal examination.

On auscultation Left sided breathe sounds were diminished in the infra-axillary area and infra-scapular area with fine end inspiratory crepts on both sides.

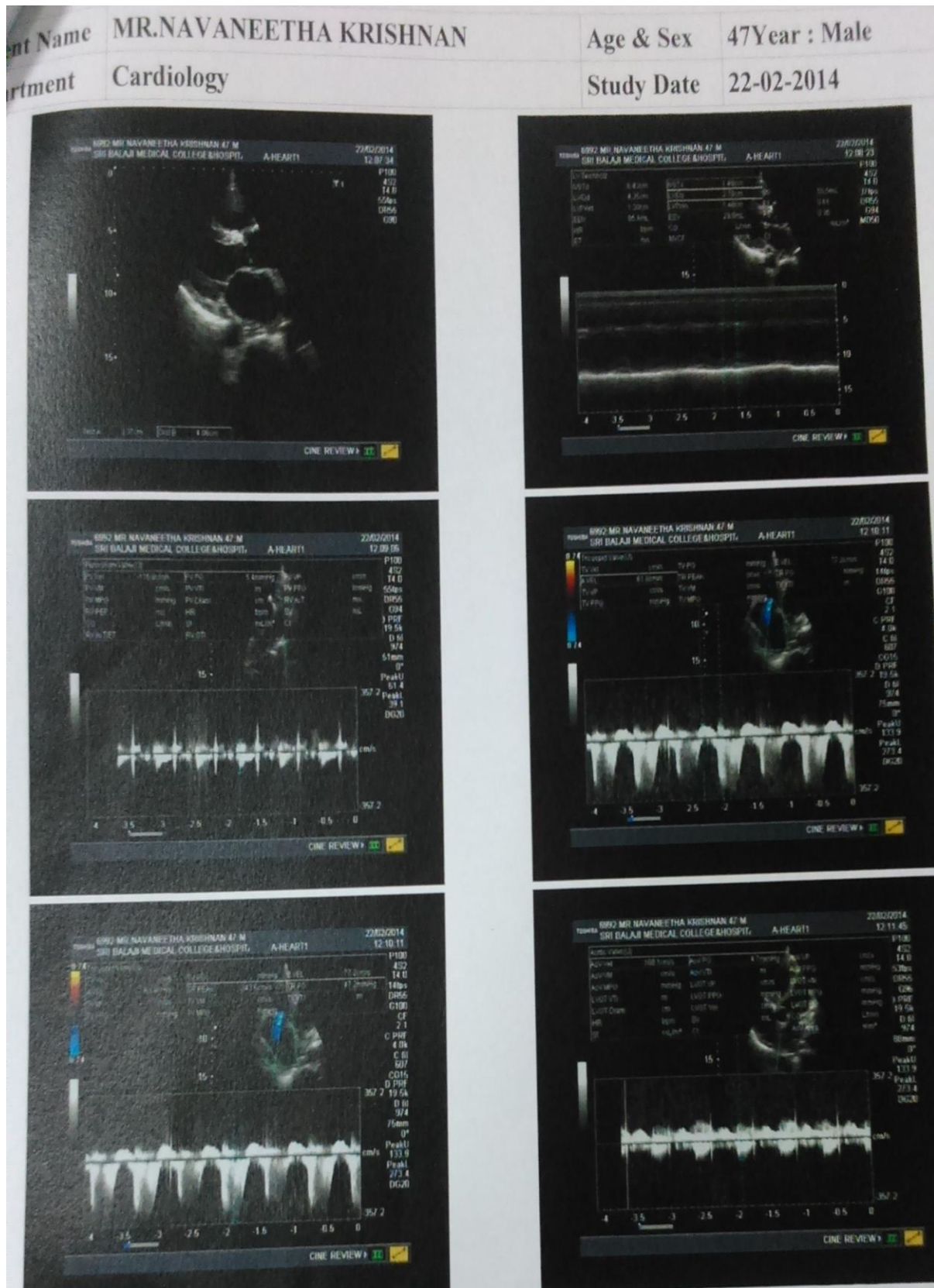
Blood investigations showed increase in blood sugar levels (FBS-127 mg/dl) and (PPBS- 231 mg/dl).

Protein electrophoresis showed monoclonal M protein >1.5g/dl.

USG abdomen showed features suggestive of hepato-splenomegaly.

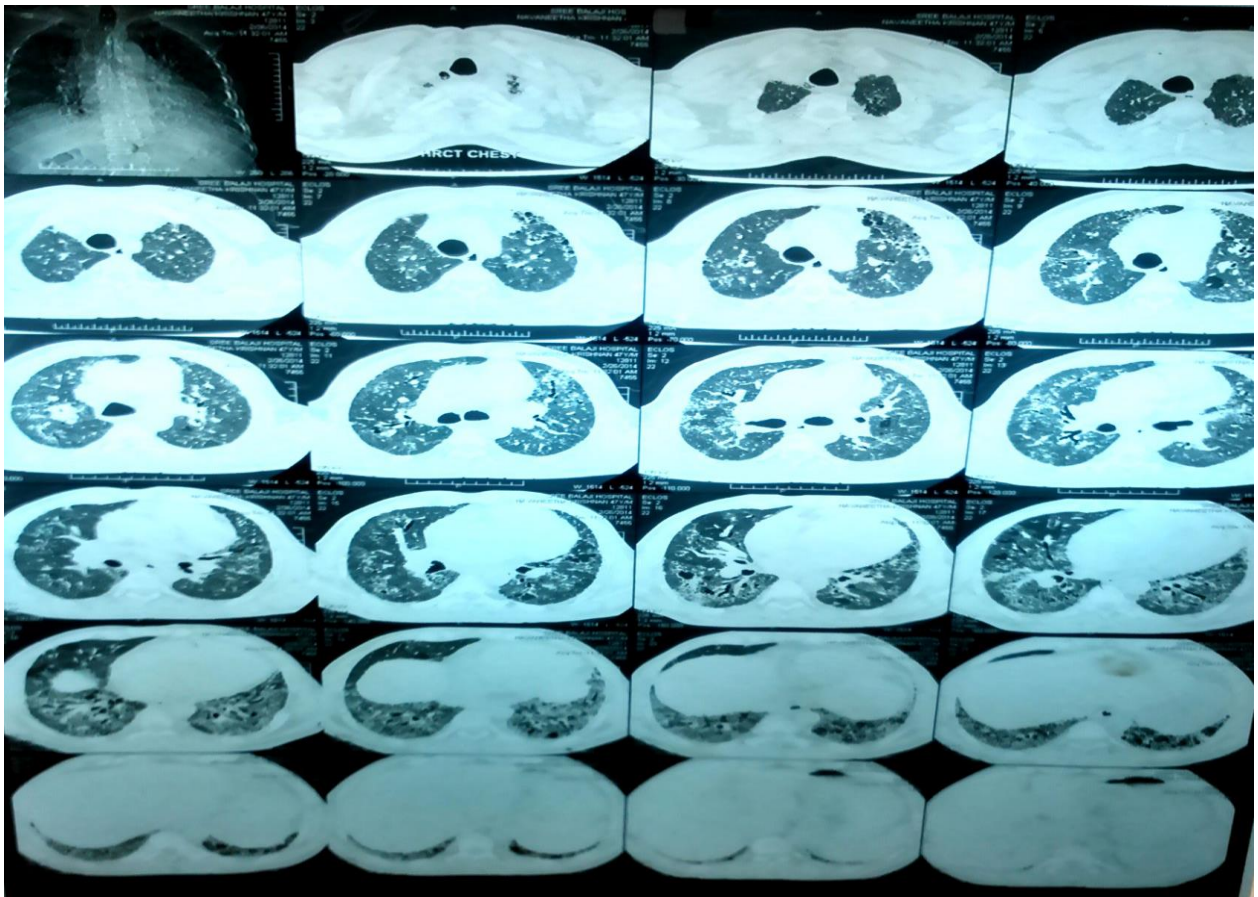
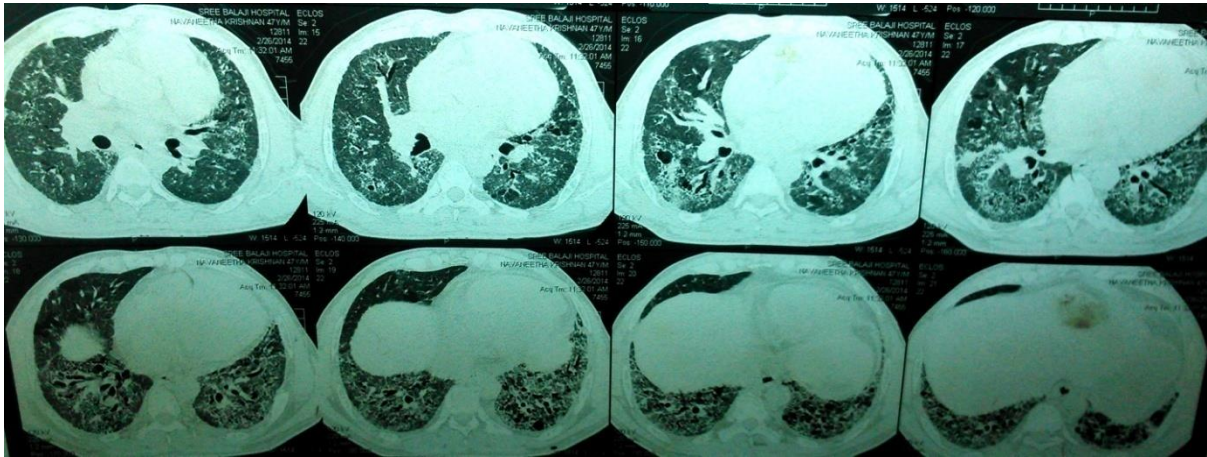


Echo Showed evidences of Moderate Pulmonary hypertension (Pulmonary flow-1.15m/sec) and minimal pericardial effusion.





CT-Chest Showed evidences of Bilateral reticular opacities with subpleural fibrosis, Traction Bronchiectasis and honey-combing of bilateral lung fields predominantly bilateral lower lobes, which were indicative of features suggestive of Interstitial Lung disease.



The presence of hepato-splenomegaly , endocrine dysfunction like Diabetes Mellitus, hypopigmentation of skin , clubbing, white nails, ascites , pleural effusion , presence of monoclonal M protein and CT –Chest showing features of Interstitial Lung Disease led to the diagnosis of Interstitial Lung Disease with Pulmonary Hypertension and POEMS syndrome.

#### DISCUSSION

Syndrome of polyneuropathy , organomegaly , endocrinopathy , monoclonal gammopathy and skin changes is a rare multi-systemic plasma cell dyscrasia.

With multiple synonyms like Crow-Fukase syndrome , Takatsuki Syndrome , multiorgan involvement , this disorder can be easily missed if index of suspicion is not very high.

The onset of POEMS Syndrome occurs most frequently in the fifth or sixth decade of life, with a mean patient age at onset of 45 years age for male and 55 years for female.[4]

The diagnosis of POEMS Syndrome requires 3 or more of 5 features and based on it our diagnosis was made.

The treatment of POEMS Syndrome depends on the treatment of underlying plasma cell disorder. Most of the patients are treated with a combination of cortico-steroids, low dose alkylators and peripheral blood stem cell transplantation following high dose chemotherapy.[2][4]

### CONCLUSION

POEMS Syndrome is a distinct pathological entity with diverse clinical manifestation.

In our patient 4 of the main 5 components of the syndrome were present.

Pulmonary Hypertension is a common feature of POEMS Syndrome and is associated with signs of extra-vascular volume overload.[3]

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