Systemic sclerosis with Usual Interstitial Pneumonia

KARTHIKEYAN

Department of General Medicine, COIMBATORE MEDICAL COLLEGE

Abstract: Systemic sclerosis is a connective tissue disorder of unknown etiology, heterogeneous clinical manifestations, and chronic and often progressive course. Pulmonary involvement is the leading cause of death in this condition. Although pathogenesis of this disease is well studied and mortality due to renal crisis declined with ACE inhibitor therapy, effective treatment strategy for Interstitial Lung Disease is yet to be found [1]. Among different types of lung involvement in systemic sclerosis, Usual Interstitial Pneumonia pattern is uncommon and carries poor prognosis. We present one such case of progressive systemic sclerosis with Usual Interstitial Pneumonia.

Keyword: systemic sclerosis, usual interstitial pneumonia

Case Report

A 45-year-old female from Ooty was admitted in our ward with complaints of cough and progressive breathlessness for the past 2 years. She had a history of bluish discoloration of fingers and toes for 5 years and digital ulcers for 3 years. She had left her job in a Tea estate 5 years back because of pain and stiffness over all major joints and difficulty in walking. Her bladder and bowel habits were normal though she had difficulty in swallowing for the past 1 year. She had two children with uneventful pregnancy and delivery. There was no significant medical history in the past.

On examination she was ill nourished with expressionless face and the skin was taught with loss of hair all over the body. We could notice Raynaud’s phenomenon [fig3], digital ulcers over fingers and toes and hyperpigmentation [fig1]. Acro-osteolysis [fig2] was present in her right hand. Pulses could be felt in all palpable peripheral vessels.

Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of cardiovascular system showed loud P2 over the second aortic area. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

On examination she was ill nourished with expressionless face and the skin was taught with loss of hair all over the body. We could notice Raynaud’s phenomenon [fig3], digital ulcers over fingers and toes and hyperpigmentation [fig1]. Acro-osteolysis [fig2] was present in her right hand. Pulses could be felt in all palpable peripheral vessels.

Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of cardiovascular system showed loud P2 over the second aortic area. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.

Her routine blood investigations revealed micromicrocytic hypochromic anemia with hemoglobin of 10gm% and normal renal, liver and thyroid function tests. Urine analysis was normal with 24 hours urine protein estimation of 20mg.

On cardiovascular system examination, there was loud P2 and grade 3 ejection systolic murmur in pulmonary area. Examination of respiratory system showed "Velcro" crackles in infra axillary and axillary areas of both hemithorax. Examination of the abdomen and nervous system were normal. We proceeded with investigations keeping in the mind the possibility of connective tissue disorder, probably systemic sclerosis.
We started cyclophosphamide pulse therapy with low dose prednisolone. Cyclophosphamide therapy, oral or intravenous, has been shown to reduce the progression in early symptomatic cases and to improve lung functions and dyspnea[2]. Steroids in systemic sclerosis do not influence the progression of internal organ involvement and when used in high doses are associated with scleroderma renal crisis[2][5][6]. Some studies showed that high dose prednisolone with cyclophosphamide to stabilize the lung function in severe ILD in systemic sclerosis.[7][8]. So, the role and the dose of steroid in this condition are controversial.

Among the various options for management of pulmonary arterial hypertension, continuous administration of intravenous prostacyclin has been found to be successful.[9] Inhaled and subcutaneous administration also provided good results. First-line bosentan therapy also has been shown to improve survival in patients with advanced PAH.[10]

**Conclusion:**

The internal organ involvement is asymptomatic till the late stage so that regular follow up with routine screening with pulmonary function tests and doppler echocardiography should be done in a case of systemic sclerosis. Therefore, ILD and pulmonary hypertension will be identified in early stage and patient will be given the benefit of early treatment.

**References:**

1. Ramnath Misra, Dhanita Khanna. Advances in Therapy of Systemic Sclerosis. JAPI 2006 ;54 ;48-51
2. Harrisons principles of internal medicine 18th edition, pages 2757-69
3. Milind Y Adakar, Nayan K Desai. Lung Involvement In Systemic Sclerosis. JAPI Medicine Update-2011;298-303

- acro osteolysis
- raynaud's phenomenon
- bilateral lower zone infiltrates
- Hyperpigmentation
- ground glass opacities and honeycombing
- microstomia