MIXED CONNECTIVE TISSUE DISEASE IN A MALE PATIENT

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ABSTRACT

A case reporting the rare occurrence of MTCD in a male patient.

Key words: MCTD, Male patient

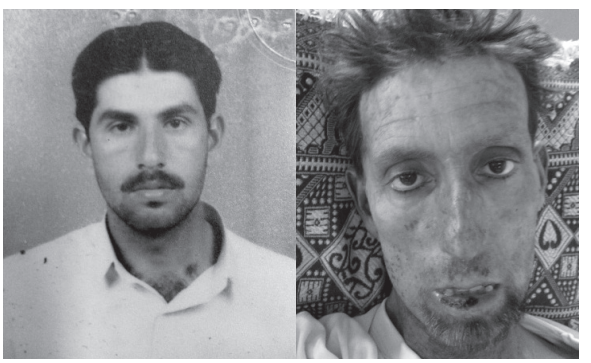
INTRODUCTION

Mixed connective tissue disorder also known as Sharps syndrome,

first recognized by Sharp and colleagues in 1972 is a disease with overlapping features of SLE, scleroderma, rheumatoid arthritis and myositis with the presence of a distinctive antibody against Anti U1-ribonucleoprotein(RNP)1,2.Completely characterized in recent years with the following core clinical and laboratory features [3,4].

- Raynauds phenomena
- Swollen hands
- Arthritis\Arthralgia
- Acrosclerosis
- Esophageal dysmotility
- Myositis
- Pulmonary hypertension

  High levels of anti-U1-RNP antibodies
- Antibodies against U1-70 kd small nuclear ribonucleoprotein



Before the onset of disease in 1997

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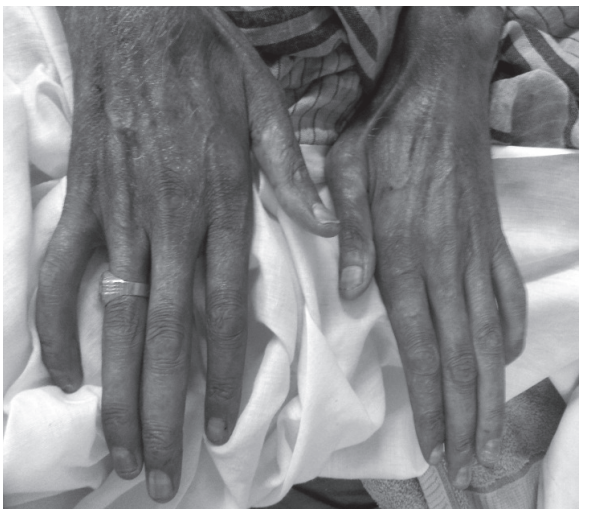
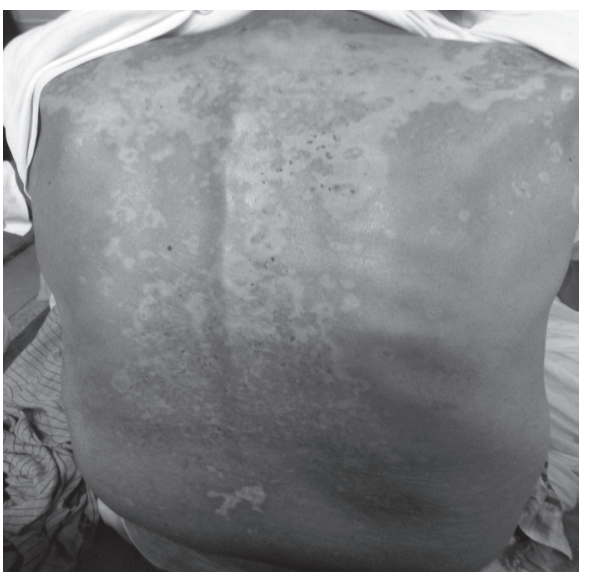




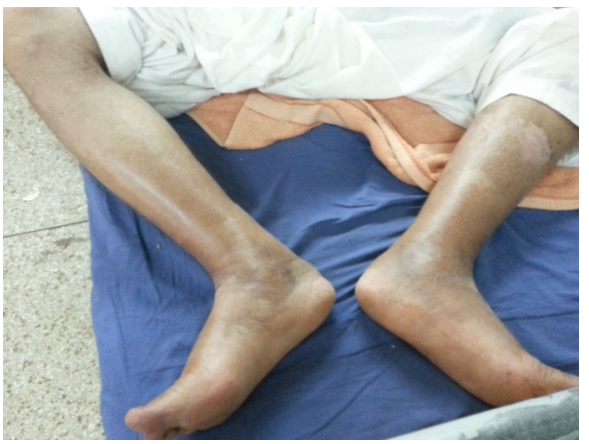
At present

CASE REPORT

Forty three years old male patient, previously admitted to medical and skin units several times and treated for polymyositis with steroids presented to us with a long standing history of low grade fever, weight loss, generalized fatigability, difficulty in swallowing food, swelling and pain in multiple small joints for the past 13 years. He recently developed severe exertional dyspnea and body swelling predominantly in the ab-

domen and lower extremities, compelling him to seek immediate medical care.

On examination he was a tall 6 ft 2" man weighing 75 kg with a marfanoid habitus. A note was made of raised JVP, multiple perioral ulcers and taut skin , decreased muscle mass , long thin upper extremities and slender fingers with gottron's papules on the knuckles , hypo pigmented skin patches on extremities along with chest and back and massive pitting edema bilaterally in the lower limbs .On lungs auscultation, diffuse bilateral coarse crackles were heard with decreased air entry at the base of the right lung whereas heart auscultation revealed added pansystolic murmur at the cardiac apex that radiated towards the axilla. Abdominal examination revealed hepatosplenomegaly.



Investigations done showed low Hb of 6.6 gms/dl ,TLC 11000 cells/cm3 ,platelet count of 60,000 .ESR of 65mm/hr, urea 142 mgs/dl ,creatinine 2.1 mgs/dl ,serum albumin 2.7mgs/dl,urine R/E +1 for albumin with numerous Rbc's and 6-8 pus cells.

24 hrs urinary collection showed proteinuria in nephritic range 225 mg/dL.CK-MB was slightly raised 32(normal=less than 25). Serum CPK serum TFT's were normal with negative VDRL test

X-ray and HRCT showed moderate right and mild left pleural effusion with large consolidation patches bilaterally.barium swallow study was normal. U/ S abdomen and pelvis showed Hepatosplenomegaly. Trans esophageal echo revealed moderate left ventricular systolic dysfunction with EF of 40-45%, anteroseptal amd Inferior wall hypokinesia moderate to severe MR moderate PAH with PAP of 52mm hg. Based on the presence of classical clinical features, Anti-U1-RNP antibodies were ordered which turned to be in significantly high titers.

DISCUSSION

MCTD is a rare condition associated with HLA-DR4 with over lapping specific features of SLE, systemic sclerosis, rheumatoid artthitisand myositis. Its prevalence appearsless than SLE (15-50 cases per 100,000 populations) and more than dermatomyositis (1-2 cases per 100,000 populations). MTCD's predilection for occurrence on female sex is well established with ratio of 3:1 [5]. The above mentioned case is one of the rare occasions when the characteristic symptoms of MCTD featured in a male patient.

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