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A Case of Undifferentiated Connective Tissue Disorder

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Abstract

Undifferentiated connective tissue diseases (UCTDs) are less common entities characterised by signs and symptoms suggestive of a systemic autoimmune disease, which do not fulfil the diagnostic criteria for a defined connective tissue disease. It is difficult to determine the prevalence of UCTD, but 10% to 20% of referrals to tertiary care clinics may be for this diagnosis. We are reporting a case without any internal organ involvement.

Key words: Connective tissue-related, undifferentiated disease, no systemic organ involvement.

Introduction

The term undifferentiated connective tissue disease (UCTD) is characterized by features suggestive of CTD which do not meet the classification criteria of the American College of Rheumatology for a specific single disease, such as systemic lupus erythematosus, systemic sclerosis, polymyositis/ dermatomyositis, and Sjögren's syndrome.[1] UCTD can evolve in these patients over time. Kinder et al.[2], had suggested prespecified criteria for diagnosis of UCTD, which are at least one of the following symptoms: Raynaud's phenomenon, arthralgias/multiple joint swelling, photosensitivity, unintentional weight loss, morning stiffness, dry mouth or dry eyes (sicca features), dysphagia, recurrent unexplained fever, gastroesophageal reflux, skin changes (rash), oral ulceration, nonandrogenic alopecia, and proximal muscle weakness; and at least one of the following indicators of systemic inflammation in the absence of infection: antinuclear antigen, rheumatoid factor, anti-SCL 70 antibody, anti-SS-A or SS-B antibody, anti-Jo-1 antibody, sedimentation rate (>2 times normal), C-reactive protein. This patient should be diagnosed and treat early to prevent their disease progression.

Case Report

A 22 years married female presented to us maculo-papular rash over cheek, upper limbs and upper trunk for 13 months. The rashes were non-itchy, no changes on sun exposure without any scar formation. There was associated multiple joint pain of hand and foot for one year. There was no significant morning stiffness. There was no history of fever, oral ulcer, joint swelling,

raynaud's phenomenon or pregnancy loss. On examination she had mild pallor, there was butterfly shaped malar rash and maculo-papular rash over both upper limb and upper trunk. There was mild tenderness in hand and foot joints and both elbow joints. But there was no other joint tenderness, swelling or any deformity. Other system examinations were all normal. Her routine blood examination showed hemoglobin 10.8 mg/dl with normocytic normochromic anemia and ESR is 30. All blood biochemistry and CRP are normal. Her routine urine examination did not show any proteinuria. Her blood for ANA profile showed highly positive ANA with positive U1RNP and Anti Sm. Rheumatoid factor and anti-CCP antibody were normal. Serum C3, C4 normal, direct coombs test was normal. Her HRCT thorax was also normal. After a multidisciplinary discussion (dermatologist and rheumatologist) a final diagnosis of UCTD was made.

Discussion

Different Rheumatologic studies have estimated that up to 25% of patients with features of a systemic autoimmune disease do not fulfill ACR classification criteria for CTD (14–18). These patients are considered to have diffuse or undifferentiated CTD (UCTD). The majority of such cases (65–94%) after years of follow-up do not develop into a ''differentiated'' CTD (e.g., rheumatoid arthritis, systemic lupus erythematosus, systemic sclerosis, mixed CTD).[3] Consequently, it has been proposed that UCTD represents a distinct clinical entity with the following criteria: signs and symptoms suggestive of a CTD, positive serologic results, and disease duration of at least 1 year.[3]

These conditions may be difficult to distinguish from early phases of defined diseases such as systemic lupus erythematosus, systemic sclerosis and others. The natural history of these rare entities is variable: a high percentage of patients with UCTD maintain an undifferentiated clinical course and do not evolve to a distinct CTD, whereas some patients can evolve over time.[1] Laboratory test screening is essential to identify markers that may suggest a systemic, autoimmune disease, or specific organ involvement. In fact, about 90% of the patients with UCTD show positive ANA.[4,5]

The onset of UCTD is similar to most CTDs, peaking in the middle years of life. There are no specific signs or symptoms of UCTDs because these entities present manifestations common to other CTDs. Major organ involvement is unusual and the lung has been reported as a late complication, often determining a worse outcome.[6]

We have described a case of a patient with UCTD whose first clinical manifestation was skin rashes. UCTD should be considered when a young female presented with prolong skin rash which may not associated with other clinical features of connective tissue diseases.

Legends of Figures

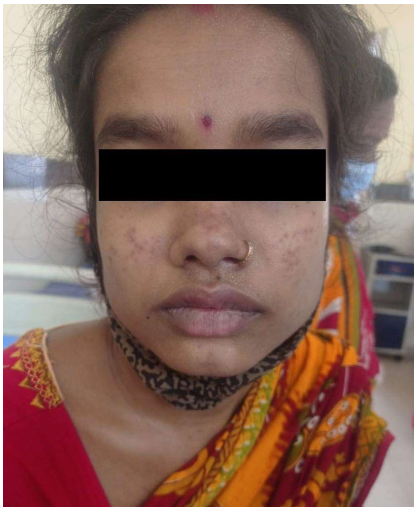

Fig 1: Butterfly lesions over face

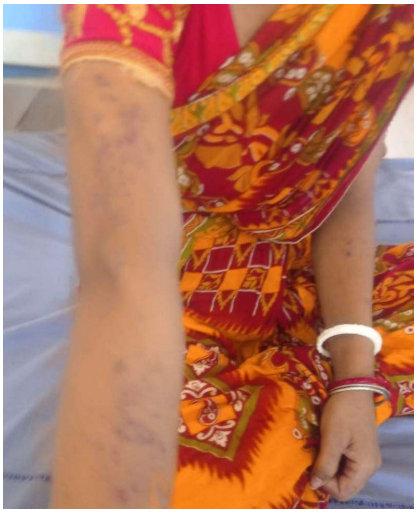

Fig 2: Maculo-papular rash on upper limb

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