CASE REPORT

Acute Diffuse Digital Gangrene- A Rare Initial Manifestation of Mixed Connective Tissue Disorder (MCTD).

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Abstract:

Introduction: Mixed connective tissue disorder (MCTD), unlike other connective tissue disorders have a milder course. MTCD with only digital gangrene without organ damage, as an initial presentation is rare. Case Report : A young female presented with c/o fever and blackish discoloration of all toes and fingers (toes > fingers). All her major organ functions were normal. Her CRP was raised and U1 snRNP antibody was positive. She responded well to injectable steroids. Conclusion: This patient with presentation of diffuse digital gangrene was diagnosed with MTCD, without major organ involvement and showed good recovery with steroids. So MCTD is a reversible cause of digital gangrene and should be considered in young females without risk factors of developing gangrene.

Key Words : Digital gangrene, MCTD, U1 snRNP antibody, Steroids.

Introduction:

A digital gangrene is an ischaemic phenomenon in which there is decreased or absent blood supply sufficient to cause necrosis of digits. There are vast variety of causes for peripheral arterial diseases. MCTD is one of them [1]. MCTD is an autoimmune disorder with presence of distinctive antibody against U1-snRNP with overlapping features of Systemic lupus Erythematous (SLE), scleroderma, and myositis [2.3]. Here we came across a MCTD with gangrene of multiple digits of upper and lower limb without other menifestations of the disease .This is been reported so as to stress the fact that MTCD may present as cutaneous vasculitis in the form of digital gangrene and it is one of the reversible cause of digital gangrene if detected earlier, and do well with injectable steroids .

Case Report:

A 25-year-old female presented to Dhiraj Hospital with complaints of black discoloration of all toes and fingers (toes > fingers) since 15 days (Fig 1) and fever since 7 days. No associated h/o bluish discolouration of extremities on cold exposure/ headache/ difficulty in swallowing/ decreased urine output. On examination, pallor was present; blackening of all toes and finger of upper limb was seen (Figure 2 and 3).

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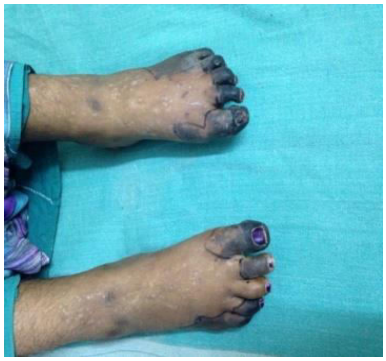
Peripheral pulses (dorsalis pedes) were normal. Examination of other organ systems was normal. On investigation, Hemoglobin was 8.7 gm% (MCV-71.4). Rest of the reports (RFT, LFT, Electrolytes, Urine, Serology, PT, aPTT) were all normal. Sickling or RA factor was negative. Reticulocyte count was 0.5%, TSH was 4.2, CRP was 145.1. Bilateral lower limb arterial Doppler study was normal. Antinuclear antibody (ANA) test was strongly positive and ANA profile showed "Anti U1 snRNP antibody". Based on these tests, patient was diagnosed as a case of Mixed Connective Tissue Disorder. She was put on injectable methylprednisolone and cyclophosphamide pulse therapy. She became symptomatically better with clinical improvement in the line of demarcation of gangrene.



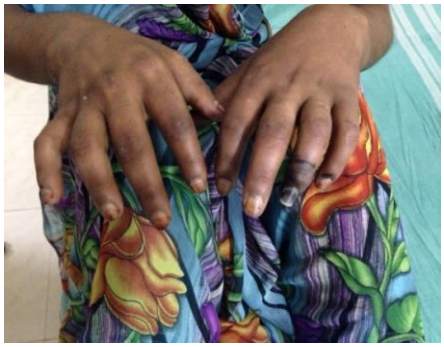


Figure 1: ***Figure 2:*** ***Figure 3 :***

Figure 1 : Patient presented with features of Diffuse digital gangrene
Figure 2 : Digital gangrene in toes of bilateral lower limb
Figure 3 : Digital gangrene of fingers of bilateral upper limb

Discussion:

MCTD also known as Sharp's syndrome was recognized by Sharp and colleagues in 1972, as an autoimmune disorder with presence of distinctive antibody against U1-RNP with overlapping features of Systemic lupus Erythematous (SLE), scleroderma, and myositis [1,2]. Therefore it is an undifferentiated connective tissue disease. It commonly causes joint swelling, malaise, Raynaud's phenomenon (90% of cases), Sjogren's syndrome, muscle inflammation and sclerodactyly. Unlike other connective tissue disorders, it has a milder course.Distinguishing lab characteristics are positive speckled ANA and an anti U1-snRNP antibody. It is associated with HLA DR-4 [4].

Based on factors like female sex, young age, no clinical evidence suggestive of alternate etiology for digital gangrene and raised CRP, patient was suspected to have some autoimmune phenomenon as causative factor and subjected for further investigation in form of ANA testing. Multiple digital gangrene is known to be one of the manifestation of MCTD, but usually it occurs in later stages of the disease along with other common menifestations as stated above. Digital gangrene as initial and first presentation is a rare phenomenon and required high suspicion to diagnose the condition.

Vasculitis induced injury to blood vessel may lead to increased vascular permeability, vessel weakening that cause aneurysm formation, haemorrhage and intimal proliferation, thrombosis that result in obstruction and local ischemia. It is critical to distinguish vasculitis occurring as a primary autoimmune disorder from vasculitis occurring secondary to infection, drugs, malignancy or connective tissue disease such as systemic lupus erythematous or rheumatoid arthritis [5,6].

Immunoglobulin finding may be helpful in diagnosing MCTD. Indirect Immuno fluroscent test – Presence high titre of IgG against U1RNP is the only autoantibody supports a diagnosis of MTCD [7]. Treatment option includes corticosteroids, Immunosuppressive drugs like cyclophosphamide (in severe vasculitis) to reduce the inflammation. IV cyclophosphamide is better compared to oral form.8Our patient responded well to steroids and cyclophosphamide pulse therapy.

Newer treatment approach includes deoxysperagualin, achieved a high rate of disease remission and permitted prednisolone reduction. Other newer Immunosuppresives are Leflunamide, TNF antagonists – Infliximab and ENBREL [9].

Conclusion:

MCTD confined to skin in form of acute diffuse digital gangrene and without any major organ involvement is rare presentation. If diagnosed earlier then they respond well to treatment with steroids and has good prognosis. So Mixed Connective Tissue Disorder is a reversible cause of digital gangrene.

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