ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 05 | May - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

DIGITAL GANGRENE – SOLE MANIFESTATION OF MIXED CONNECTIVE TISSUE DISORDER: A CASE REPORT

Internal Medicine

Dr. M. Mathur Professor, Department of Internal Medicine, MLN Medical College, Prayagraj

Dr. Aman Yadav 3rd year resident, Department of Internal Medicine, MLN Medical College, Prayagraj

Dr. Deepanshu Dhaneshary 3rd year resident, Department of Internal Medicine, MLN Medical College, Prayagraj

ABSTRACT

It is unusual to see MTCD with only digital gangrene as the first presentation, with no other systemic symptoms. We hereby report the case of a young female presented with c/o blackish discoloration of fingers of the upper limb sparing thumb. All of her main organs functioned normally. Her CRP was elevated, and U1 snRNP antibody was positive. This patient presented with widespread digital gangrene and was diagnosed with MTCD, which did not impact major organs. As a result, MCTD is a cause of digital gangrene and should be examined in young females who do not have other risk factors for the disease.

KEYWORDS

Digital Gangrene; MCTD; Female; U1 snRNP antibody; Sharp's syndrome

INTRODUCTION

A digital gangrene is an ischaemic phenomenon in which there is a decreased or absent blood supply, resulting in necrosis of the digits. There are several causes of peripheral arterial disease. One such condition is mixed connective tissue disease (MCTD) . MCTD is a rare autoimmune disease. MCTD, as first described in 1972, is "classically" characterized by overlapping features of various systemic connective tissue diseases, including systemic lupus erythematosus, systemic sclerosis, dermato-/polymyositis, and rheumatoid arthritis. They also exhibit extremely high levels of antinuclear antibodies (ANAs) and antibodies to ribonucleoprotein (anti-RNP) in their blood .

Diagnosis can be challenging due to variable and diverse symptoms upon presentation and the changes in symptoms over time. Alarcon-Segovia is one of the regularly used diagnostic criteria that consists of a high titer of positive anti-U1-RNP (over 1 per 1600) and three or more of the following clinical manifestations: Raynaud phenomenon, hand edema, synovitis, histologically proven myositis, and acrosclerosis . The patient described here showed many of the characteristics of this syndrome, a distinctive feature being the severity of Raynaud's phenomenon, ultimately leading to gangrene of multiple digits of the upper limb without other manifestations of the disease.

Case Report

The patient (Annu Devi), a housewife, presented to Swaroop Rani Nehru Hospital with complaints of black discoloration of the fingers of both upper limbs (sparing thumbs of the upper limb) for 3–4 months. There was no associated history of bluish discoloration of fingers on cold exposure, nor was there any difficulty in swallowing or frothing urine. Pallor was apparent on examination, along with dry gangrene of the upper limbs, except the thumb. The rest of the systemic examination was within normal limits. Her pulse rate was 88/min, regular, normovolemic and all peripheral pulses were palpable while blood pressure was 110/70 mm Hg. There was digital gangrene involving the fingers (Figure 1).

Figure 1: Dry gangrene in finger tips of all upper limbs, except thumbs.

Investigations

Biochemical tests showed haemoglobin of 8.7 g/mL (MCV-71.4). The remainder of the reports (RFT, LFT, electrolytes, urine, serology, PT, and aPTT) were all normal. The RA factor was negative. The reticulocyte count was 0.5%, TSH was 4.2, and CRP was reactive. The bilateral lower-limb arterial Doppler study was normal.

Immunologic studies like antinuclear antibody (ANA) was performed, with a strongly positive homogeneous pattern. Based on these tests, the patient was diagnosed as having MCTD.

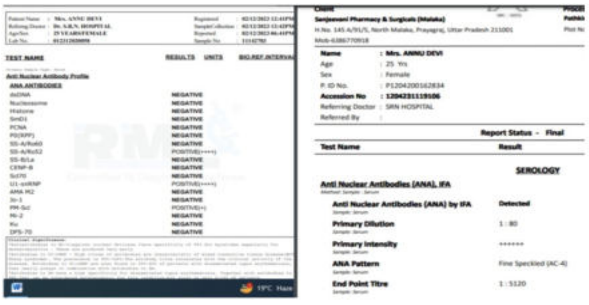

Figure 2: Investigations.

DISCUSSION

MCTD, also known as Sharp's syndrome, was recognised by Sharp and colleagues in 1972 as an autoimmune disorder with a distinctive antibody against U1-RNP and overlapping features of systemic lupus erythematous (SLE), scleroderma, and myositis. The U1-RNP complex is an intranuclear protein that converts pre-mRNA to mature RNA. It constitutes three specific proteins, A, C, and 70 kDa, to which anti-U1 RNP antibodies bind. The 70 kDa antigen is the main target of the anti-RNP antibody in MCTD. Environmental factors, e.g., infections, drugs, toxins, UV radiation, and chemicals, including vinyl chloride and silica, have correlations with the development of MCTD . Initial symptoms of mixed connective tissue disease are usually non-specific and include arthralgia, malaise, myalgia, and low-grade fever. Almost any organ system can be affected by MCTD.

The most common skin change is the Raynaud phenomenon, which is also the most common presenting feature of MCTD. Hand oedema, acrosclerosis, calcinosis cutis, telangiectasia, erythema nodosum, hair loss, and vasculitis of the digits are other manifestations. Discoid plaques and a malar rash similar to SLE can also occur. Nailfold changes are visible by capillary microscopy. Case reports describe panniculitis involving the abdomen, legs, and breasts [5].

Multiple digital gangrene is known to be one of the manifestations of MCTD, but usually it occurs in later stages of the disease along with other common manifestations, as stated above.

CONCLUSION

MCTD confined to the skin and presenting only as gangrene without systemic involvement is rare. Keeping digital gangrene as one of the

initial manifestations of MCTD can help in the early diagnosis of the disease.

REFERENCES

1. Sharp GC, Irvin WS, Tan EM, Gould RG, Holman HR. Mixed connective tissue disease-an apparently distinct rheumatic disease syndrome associated with a specific antibody to an extractable nuclear antigen (ENA). The American journal of medicine. 1972 Feb 1; 52(2):148-59.
2. Alarcon-Segovia D, Cardiel MH. Comparison between 3 diagnostic criteria for mixed connective tissue disease. Study of 593 patients. The Journal of rheumatology. 1989 Mar 1;16(3):328-34.
3. Greidinger EL, Zang YJ, Jaimes K, Martinez L, Nassiri M, Hoffman RW. CD4+ T cells target epitopes residing within the RNA-binding domain of the U1-70-kDa small nuclear ribonucleoprotein autoantigen and have restricted TCR diversity in an HLA-DR4-transgenic murine model of mixed connective tissue disease. The Journal of Immunology. 2008 Jun 15;180(12):8444-54.
4. Khanna A, Suri JC, Ray A, Sharma RK. Silica associated mixed connective tissue disorder in a stone crusher. Indian Journal of Occupational and Environmental Medicine. 2013 May 1;17(2):76-7.
5. Nakashima M, Suzuki K, Okada M, Kataharada K, Takada K, Nakanishi T, Kobayashi M, Ohsuzu F. Panniculitis in a patient with mixed connective tissue disease. Modern rheumatology. 2004 Jul 1;14(3):250-3.
6. Katzen BT. Clinical diagnosis and prognosis of acute limb ischemia. Reviews in cardiovascular medicine. 2002 Jan 1;3:S2-6.