RESEARCH PAPER

Severe Digital Ischemia and Necrosis in Mixed Connective Tissue Disease: A Case of Secondary Raynaud's Phenomenon Leading to Digital Shortening

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ABSTRACT
We present the case of a **48-year-old female patient** diagnosed with **Mixed Connective Tissue Disease (MCTD)** who developed severe secondary Raynaud's phenomenon [i]. This complication resulted in critical **digital ischemia and necrosis**, specifically affecting the right ring finger, leading to digital shortening [i]. Diagnosis was strongly supported by classic serological markers, including a **positive Anti-Nuclear Antibody (ANA)** test with a **speckled pattern** [i], high titres of **Anti-Nucleosome** antibodies [i], and definitive positivity for both **RNP 68kd and Sm/RNP** antigens [i]. This report highlights the destructive potential of severe secondary Raynaud's in MCTD and the importance of timely diagnosis based on specific serological profiles.

Keywords: *Mixed Connective Tissue Disease, Raynaud's phenomenon, Digital ischemia, Anti-U1 RNP antibodies, Connective tissue disorders*
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Introduction

Mixed Connective Tissue Disease (MCTD) is a systemic autoimmune disorder characterized by clinical features overlapping those of systemic lupus erythematosus (SLE), systemic sclerosis (SSc), and polymyositis (PM) [i]. The defining feature for the diagnosis of MCTD is the presence of high titres of antibodies directed against the **U1 ribonucleoprotein (U1 RNP)** [i]. A common initial symptom, often preceding the full clinical picture by years, is Raynaud's phenomenon [i]. While mild Raynaud's is common, severe secondary Raynaud's can lead to critical digital ischemia, digital ulcers, gangrene, and eventual tissue loss or shortening of the digit [i].

Case Presentation

A 48-year-old female presented with clinical symptoms consistent with Mixed Connective Tissue Disease (MCTD) [i]. Her history included **secondary Raynaud's phenomenon**, which had progressed severely to affect the right ring finger [i]. Clinical examination revealed severe damage to the affected digit, confirming that the right ring finger was **shortened** [i]. Visual inspection of the hands showed areas of skin changes, possibly indicative of sclerodactyly or healed lesions. The **right ring finger exhibited severe digital necrosis/gangrene**, appearing blackened and damaged.

Laboratory investigations confirmed the autoimmune nature of the disease:

1. **Anti-Nuclear Antibody (ANA) testing** was **positive** [i].
2. The immunofluorescence pattern displayed a **speckled pattern** [i].
3. Specific autoantibodies were highly indicative of MCTD, including positivity for **Anti-Nucleosome** antibodies [i].
4. Crucially, the patient was **positive for RNP 68kd** [i].

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5. Further antigen testing demonstrated positivity for **Sm/RNP** [i].
These clinical and serological findings confirmed the diagnosis of Mixed Connective Tissue Disease with severe vascular complications [i].

Diagnosis

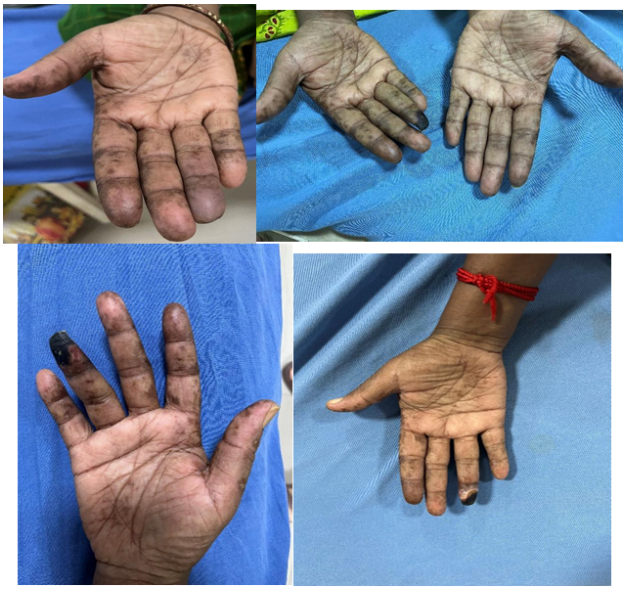
The diagnosis of MCTD was confirmed based on the coexistence of characteristic clinical features (including severe Raynaud's phenomenon leading to digital damage/shortening) and the pathognomonic serological profile [i].
The criteria supporting the diagnosis included:
1. **Clinical Manifestation:** Presence of secondary Raynaud's phenomenon resulting in digital necrosis and shortening [i].
2. **Serological Hallmark:** The definitive diagnosis hinges on the detection of high titres of anti-U1 RNP antibodies [i]. This was supported by the positive findings for **RNP 68kd** [i] and **Sm/RNP** [i] antibodies.
3. **General Autoimmunity:** General ANA positivity with a **speckled pattern** further supported the underlying systemic autoimmune process [i].
The digital necrosis observed, specifically the shortened, gangrenous right ring finger [i], represents a severe manifestation of vascular involvement common in connective tissue diseases like MCTD.

Treatment Considerations

Management of MCTD is generally guided by the dominant clinical features present in the patient [i]. Given the patient's presentation of severe secondary Raynaud's leading to digital necrosis and gangrene [i], the treatment strategy must aggressively address the underlying vasospasm and vasculopathy.
While specific treatment details for this patient are not available in the sources, the management for critical digital ischemia in MCTD typically requires a multi-faceted approach:
1. **Vasoactive Therapy:** Aggressive vasodilators are critical to improve blood flow to the affected digit. This often includes dihydropyridine calcium channel blockers (e.g., nifedipine) and, in cases of critical ischemia, intravenous prostanoids (e.g., iloprost) to induce maximal vasodilation [i].
2. **Immunosuppression:** Given the underlying autoimmune etiology (MCTD), immunosuppressive agents may be employed to control systemic inflammation and prevent further vascular damage. This may involve corticosteroids or other immunosuppressants tailored to the extent of internal organ involvement, if any [i].
3. **Management of Necrosis:** The presence of established gangrene and digital shortening necessitates careful wound care to prevent infection and promote demarcation [i]. Surgical intervention, such as debridement or potential amputation of the non-viable tissue (the shortened, necrotic right ring finger) [i], would be required once demarcation is complete, or if infection supervenes.

CONCLUSION

This case illustrates a severe vascular complication—**digital gangrene and shortening of the right ring finger**—in a 48-year-old female patient with established **Mixed Connective Tissue Disease** [i]. The strong positive serology for **ANA (speckled pattern)**, **nucleosome**, **RNP 68kd**, and **Sm/RNP** provides robust evidence for the diagnosis [i]. Severe Raynaud's phenomenon leading to irreversible tissue damage underscores the need for continuous vigilance and aggressive vascular intervention in patients presenting with MCTD.



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