E-ISSN:2320-8686
P-ISSN:2321-127X
RNI:MPENG/2017/74037
Case Report
Upper-Gastro

International Journal of Medical Research and Review
2025 Volume 13 Number 2 Apr-Jun

Publisher
MEDRESEARCH
www.medresearch.in

CREST Syndrome: A Rare Cause of Chronic Upper-Gastro Intestinal Haemorrhage in Adults

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DOI:https://doi.org/10.17511/ijmrr.2025.i02.04

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CREST syndrome, a limited cutaneous variant of systemic sclerosis, rarely presents with chronic upper gastrointestinal haemorrhage. This case report explores the complex pathological mechanisms and management challenges of this uncommon manifestation. A 52-year-old female with established CREST syndrome presented with recurrent melena and hematemesis. Comprehensive clinical evaluation revealed extensive gastrointestinal telangiectasia with significant bleeding potential. Upper endoscopy demonstrated multiple fragile vascular lesions throughout the esophagus, stomach, and duodenum. The patient underwent detailed diagnostic imaging, including CT angiography and capsule endoscopy. Therapeutic interventions included argon plasma coagulation, tranexamic acid therapy, and systemic immunosuppression with mycophenolate mofetil.

Keywords: Systemic Sclerosis, CREST Syndrome, Gastrointestinal Haemorrhage, Telangiectasia, Immunosuppressive Agents

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How to Cite this Article

Sj Sanketh, Charantimath NC, Ugaragol P, CREST Syndrome: A Rare Cause of Chronic Upper-Gastro Intestinal Haemorrhage in Adults. Int J Med Res Rev. 2025;13(2):16-19.
Available From
https://ijmrr.medresearch.in/index.php/ijmrr/article/view/1537

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Manuscript Received	Review Round 1	Review Round 2	Review Round 3	Accepted
2025-03-05	2025-02-13	2025-02-21	2025-03-01	2025-03-09

Conflict of Interest	Funding	Ethical Approval	Plagiarism X-checker	Note
None	Nil	Yes	11.16	

Introduction

Systemic sclerosis (SSc), also known as scleroderma, is a complex autoimmune connective tissue disorder characterized by progressive fibrosis of the skin and internal organs. CREST syndrome, a limited cutaneous variant of systemic sclerosis, presents unique clinical manifestations that can lead to significant complications, including gastrointestinal involvement [1]. While the global SSc incidence and newlydiagnosedpopulation were estimated to be 8.64 per 100,000 person-years (1.78–23.57) and0.67 million (0.14–1.84)people annually, respectively, gastrointestinal bleeding remains an uncommon yet potentially life-threatening manifestation [2].

Chronic upper gastrointestinal haemorrhage in CREST syndrome patients can result from various pathological mechanisms, including mucosal atrophy, telangiectasia, and vascular fragility [3]. These complications arise from the underlying autoimmune process that causes widespread microvascular damage and fibrotic changes in the gastrointestinal tract [4]. Despite the recognized potential for gastrointestinal involvement, chronic upper gastrointestinal haemorrhage as a primary presentation remains a rare clinical scenario that challenges diagnostic & management strategies [5].

This case report aims to highlight the complex interplay between CREST syndrome and chronic upper gastrointestinal haemorrhage, contributing to the understanding of this unusual clinical manifestation and potentially guiding future diagnostic and therapeutic approaches.

Case Report

A 52-year-old female presented to our General Medicine OPD with a two-year history of intermittent melena and recurrent episodes of hematemesis. Her medical history was remarkable for CREST syndrome, diagnosed six years prior, characterized by calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia.

Physical examination revealed characteristic skin thickening of the hands and face, with multiple telangiectatic lesions on the lips and fingers. Laboratory investigations showed mild anaemia with hemoglobin of 9.4 g/dL, consistent with chronic blood loss.

Upper endoscopy demonstrated extensive mucosal telangiectasia throughout the esophagus, stomach, and duodenum, with multiple fragile vascular lesions prone to spontaneous bleeding.

Detailed imaging, including CT angiography and capsule endoscopy, confirmed widespread gastrointestinal microvasculature abnormalities characteristic of systemic sclerosis. The patient underwent multiple therapeutic interventions, including argon plasma coagulation and tranexamic acid therapy to manage chronic haemorrhage.

Over a six-month follow-up period, the patient received periodic transfusions and underwent aggressive management of her gastrointestinal vascular lesions. Immunosuppressive therapy with mycophenolate mofetil was initiated to control underlying autoimmune processes and potentially reduce vascular fragility.

The case highlights the complex management challenges of gastrointestinal complications in CREST syndrome, demonstrating the critical importance of a multidisciplinary approach involving rheumatology, gastroenterology, and interventional specialists in managing this rare manifestation of systemic sclerosis.

Discussion

The presented case illustrates the complex gastrointestinal manifestations of CREST syndrome, particularly focusing on chronic upper gastrointestinal haemorrhage. Our findings align with previous literature documenting the significant vascular pathology associated with systemic sclerosis, characterized by widespread telangiectasia and microvascular abnormalities [6].

Petcu A et al. reported similar endoscopic findings in a comprehensive study of 79 scleroderma patients, noting that gastrointestinal involvement occurs in up to 72.1% of cases, with significant hemorrhagic potential [7]. Our case demonstrates the extreme end of this spectrum, where chronic bleeding becomes the predominant clinical manifestation, a scenario reported in less than 5% of systemic sclerosis cases.

The management approach in our case, combining argon plasma coagulation and systemic immunosuppression, mirrors recommendations by Sallam and McNearney,

Who emphasized the importance of multidisciplinary intervention in managing gastrointestinal complications of systemic sclerosis [8]. The use of mycophenolate mofetil represents an emerging therapeutic strategy aimed at controlling both inflammatory processes and vascular fragility.

Pathophysiologically, our findings support existing literature highlighting the role of endothelial dysfunction and fibrotic changes in scleroderma-related gastrointestinal bleeding.[9]Ren H et.al. underscore the complex interplay between autoimmune processes, microvascular damage, and progressive tissue remodelling that characterizes this syndrome [10].

Comparative analysis with similar case reports reveals the unique challenges in managing gastrointestinal bleeding in CREST syndrome. While most studies report intermittent bleeding, our case demonstrates a more severe, chronic hemorrhagic pattern requiring aggressive therapeutic intervention. This underscores the heterogeneous nature of systemic sclerosis and the importance of individualized management approaches.

The long-term prognosis remains guarded, consistent with existing literature suggesting that gastrointestinal involvement significantly impacts quality of life and patient outcomes in systemic sclerosis [5,11]. Our case contributes to the growing body of evidence highlighting the need for comprehensive, multidisciplinary management strategies in these complex autoimmune conditions.

Conclusion

This case report highlights the rare but significant manifestation of chronic upper gastrointestinal hemorrhage in CREST syndrome, demonstrating the complex interplay between systemic sclerosis and gastrointestinal vascular pathology. Our findings emphasize the critical importance of comprehensive multidisciplinary management, aggressive endoscopic intervention, and targeted immunosuppressive therapy in mitigating potentially life-threatening complications. The case underscores the need for heightened clinical awareness of gastrointestinal manifestations in systemic sclerosis, particularly in patients presenting with persistent bleeding and underlying autoimmune conditions, ultimately contributing to a more nuanced understanding of this challenging clinical scenario.

Conflict Of Rights

No conflict of rights.

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