Cureus
Part of SPRINGER NATURE

Open Access Case Report

Gastric Variceal Hemorrhage in a Patient With Sclerodactyly: An Unexpected Cause of Hematemesis in Limited Cutaneous Systemic Sclerosis

Review began 09/27/2026
Review ended 11/05/2026
Published 11/09/2026

© **Copyright** 2026
Khemchandani. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

DOI: 10.7759/cureus.119364

Nidhesh Khemchandani [1]

1. Department of Gastroenterology and Hepatology, Sri Aurobindo medical College and Post Graduate Institute, Indore, IND

Corresponding author: Nidhesh Khemchandani, nidheshkhem@gmail.com

Abstract

Microvascular injury, immunological dysregulation, and increasing collagen fibrosis are the hallmarks of systemic sclerosis (SSc), an inflammatory connective tissue disease. Its limited cutaneous subset, historically termed calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia syndrome, is characterized by the presence of anticentromere antibodies. Gastrointestinal involvement is almost universal, and when overt bleeding occurs, it is usually attributed to gastric antral vascular ectasia (GAVE) or mucosal telangiectasia.

This article describes a 66-year-old woman with limited cutaneous SSc who presented with acute upper gastrointestinal bleeding due to actively bleeding gastric varices associated with portal hypertension, chronic liver disease, and extensive portal-vein thrombosis. Her physical examination revealed sclerodactyly, salt-and-pepper pigmentation, restricted mouth opening, and moderate splenomegaly, along with Raynaud phenomenon. Antinuclear antibody was positive at a titer of 1:100 in a centromere (AC-3) pattern, with anticentromere protein B reactivity, and a skin biopsy was performed. A diagnosis of SSc was made on the basis of clinical, serological, and skin biopsy findings.

Urgent esophagogastroduodenoscopy demonstrated small esophageal varices and large gastric varices that were actively bleeding and were managed with 1 mL of cyanoacrylate glue. Triple-phase computed tomography showed a cirrhotic liver, partially occlusive portal vein thrombosis, moderate splenomegaly, and a large gastrorenal shunt. Even though the etiology of liver disease was unknown, a raised serum immunoglobulin G level supported coexisting autoimmune liver disease. Secondary prophylaxis was achieved by retrograde transvenous obliteration of the gastrorenal shunt with plug assistance employing an Amplatzer vascular plug (Abbott Medical, Plymouth, MN). The patient remained free of rebleeding with a stable hemoglobin level at two and six weeks. This case illustrates that hematemesis in SSc must not be assumed to be a scleroderma-specific mucosal lesion such as GAVE or mucosal telangiectasia, that coexisting autoimmune liver disease and portal hypertension should be actively excluded, and that endovascular shunt obliteration is an effective secondary-prophylaxis strategy.

Categories: Rheumatology, Internal Medicine, Gastroenterology
Keywords: anticentromere antibody, autoimmune liver disease, crest syndrome, gastric antral vascular ectasia, gastric varices, limited cutaneous systemic sclerosis, plug-assisted retrograde transvenous obliteration, portal vein thrombosis, upper gastrointestinal bleeding

Introduction

Systemic sclerosis (SSc) is an autoimmune connective tissue disease characterized by the triad of microvascular injury, immune activation, and progressive fibrosis of the skin and internal organs [1]. The limited cutaneous SSc was historically described by the acronym CREST (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) and is serologically characterized by anticentromere antibodies, directed principally against centromere protein B (CENP-B) [2]. These autoantibodies are highly specific for the limited subset and identify a distinct clinical phenotype.

The gastrointestinal tract is the internal organ system most frequently affected in SSc. The involvement of the gastrointestinal tract may extend from the esophagus to the anorectum. There is disproportionate involvement of the upper gastrointestinal tract, with esophageal involvement seen in up to 90% of patients in the form of reflux esophagitis, stricture formation, Barrett esophagus, and adenocarcinoma as a result of a hypotensive lower esophageal sphincter and reduced/absent peristalsis [3,4]. Overt gastrointestinal hemorrhage is comparatively uncommon, and when it occurs, it is most often attributed to two scleroderma-specific mucosal lesions: gastric antral vascular ectasia (GAVE, or watermelon stomach) and diffuse mucosal telangiectasia [5,6]. GAVE in particular has become the archetypal cause of bleeding in this population, reported in approximately 6% of patients with SSc [7].

How to cite this article
Khemchandani N (November 09, 2026) Gastric Variceal Hemorrhage in a Patient With Sclerodactyly: An Unexpected Cause of Hematemesis in Limited Cutaneous Systemic Sclerosis. Cureus 18(11): e119364. DOI 10.7759/cureus.119364

This diagnostic anchoring can be misleading. SSc and anticentromere positivity, in particular, carry a well-recognized association with autoimmune liver disease, most notably primary biliary cholangitis, the combination of which with limited cutaneous SSc is termed Reynolds syndrome [8,9]. Primary biliary cholangitis-related autoantibodies are detectable in a substantial minority of unselected patients with SSc [10]. Such patients may progress to cirrhosis, portal hypertension and variceal hemorrhage, a mechanism entirely distinct from scleroderma vasculopathy, yet clinically indistinguishable at the bedside. We report an elderly woman with limited cutaneous SSc in whom hematemesis arose from actively bleeding gastric varices in the setting of portal vein thrombosis and chronic liver disease, and in whom definitive secondary prophylaxis was achieved by plug-assisted retrograde transvenous obliteration (PARTO).

Case Presentation

History

A 66-year-old woman presented to the emergency department with multiple blood-tinged emesis, each approximately 50-100 mL in volume, accompanied by the passage of black, tarry, foul-smelling stools. There had been no preceding retching or nonbloody vomiting. She reported no previous similar episode and denied jaundice, abdominal distension, abdominal pain, anorexia, or weight loss. There was no history of using nonsteroidal anti-inflammatory drugs, and she had never received a blood transfusion. She described long-standing painful bluish discoloration of the fingers on cold exposure during the winter months, consistent with Raynaud phenomenon.

Her comorbidities comprised type 2 diabetes mellitus, treated with metformin 500 mg twice daily; hypertension of two years' duration, treated with telmisartan 40 mg once daily; and hypothyroidism, treated with levothyroxine 62.5 µg daily. She consumed a mixed diet and had never smoked or used alcohol. There was no family history of a similar condition, and she was married with two sons.

Examination

She was conscious and alert. Her heart rate was 110 beats/minute, her blood pressure was 100/70 mmHg, and she was afebrile with a 98% oxygen saturation on room air. The jugular venous pressure was normal. There was pallor with delayed capillary refill. Cutaneous examination demonstrated multiple depigmented macules distributed over the body in a salt-and-pepper pattern, together with sclerodactyly and reduced mouth opening (Figure *1*). Abdominal examination revealed moderate splenomegaly without clinically detectable hepatomegaly or free fluid.



FIGURE 1: Cutaneous manifestations of limited cutaneous systemic sclerosis in the present patient, showing sclerodactyly with tapering of the digits and salt-and-pepper depigmentation

Laboratory investigations

Laboratory results at presentation are displayed graphically in Figure *2*. There was a normocytic, normochromic anemia with a hemoglobin of 7.2 g/dL and thrombocytopenia with a platelet count of 96 × 10^3/mm^3, in the presence of a normal coagulation profile (international normalized ratio 1.03). Renal function and transaminases were normal. Two findings deserve emphasis: the alkaline phosphatase was raised at 209 IU/L in the presence of normal bilirubin and normal transaminases, constituting a cholestatic biochemical pattern; and there was hypoalbuminemia with a low total protein. The stool guaiac test was positive, confirming gastrointestinal blood loss.

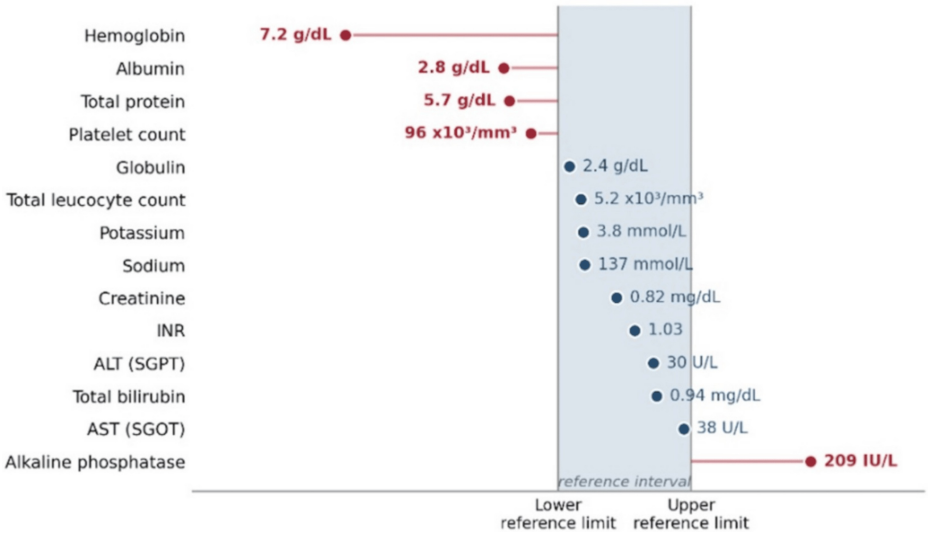


FIGURE 2: Laboratory results at presentation expressed relative to the laboratory reference interval. Each result is normalized so that the lower reference limit corresponds to 0 and the upper reference limit to 1; the shaded band therefore represents the normal range. Values falling outside the band are shown in red. The pattern of anemia, thrombocytopenia, hypoalbuminemia, and isolated alkaline phosphatase elevation is that of chronic liver disease with hypersplenism rather than of isolated gastrointestinal blood loss

INR: international normalized ratio; ALT: alanine aminotransferase; AST: aspartate aminotransferase; SGPT: serum glutamic-pyruvic transaminase; SGOT: serum glutamic-oxaloacetic transaminase

Autoimmune and serological work-up

Antinuclear antibody testing by indirect immunofluorescence was positive at a titer of 1:100 with a centromere pattern (AC-3), and the antinuclear antibody immunoblot was positive for CENP-B, supporting a limited cutaneous SSc diagnosis. Serum immunoglobulin G was raised, which, in combination with the sonographic changes of chronic liver disease and a possibility of autoimmune liver disease, was kept. Hepatitis B surface antigen, anti-hepatitis C virus antibody, and HIV viral indicators were all nonreactive. Dermatological and rheumatological opinions were obtained.

Imaging and histopathology

Changes of chronic liver disease were revealed by bedside ultrasonography. Abdominal triple-phase computed tomography showed mosaic parenchymal enhancement of the liver with irregular margins, with 60%-80% luminal narrowing due to a partially occlusive main portal vein thrombus that extends into the left and right portal veins with near-complete occlusion of the right branch; no intrahepatic biliary radicle dilatation; and moderate splenomegaly of size 9 × 14.5 × 12.5 cm (splenic index 1,631). Multiple collaterals in the splenic bed, perigastric region, and gastric wall are seen, with a large gastrorenal shunt and mild ascites, as shown in Figure *3*.

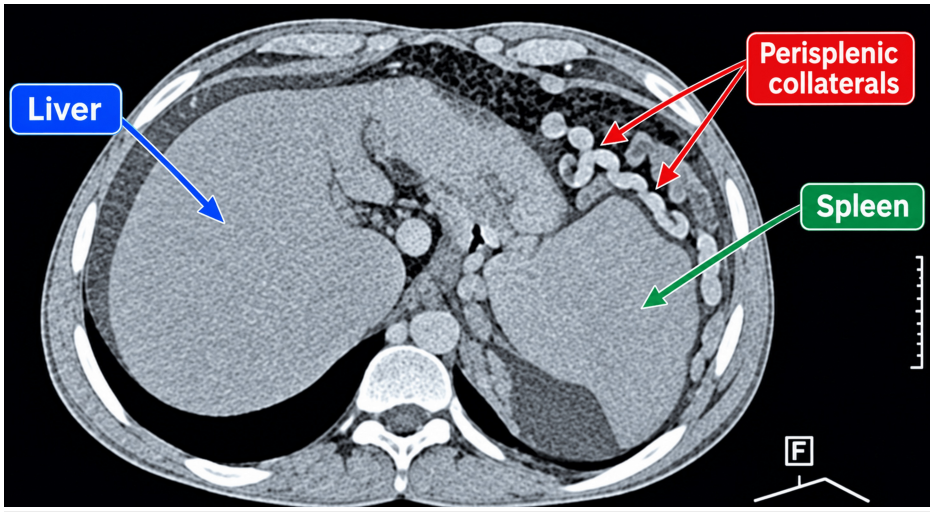


FIGURE 3: CT image showing mild ascites with splenomegaly and irregular liver margins with multiple perisplenic collaterals

CT: computed tomography

Skin biopsy (Figure *4*) revealed epidermal thinning with flattening of the rete ridges; orthokeratosis; and thinning of the granular layer, alternating hypopigmentation and hyperpigmentation with necrotic keratinocytes and vacuolar change in the basal layer, with papillary dermis showing fine sclerosis with eosinophilic material deposition. The superficial dermis contained sparse perivascular lymphocytic infiltrate and homogenized horizontal collagen bundles. The changes were consistent with SSc.

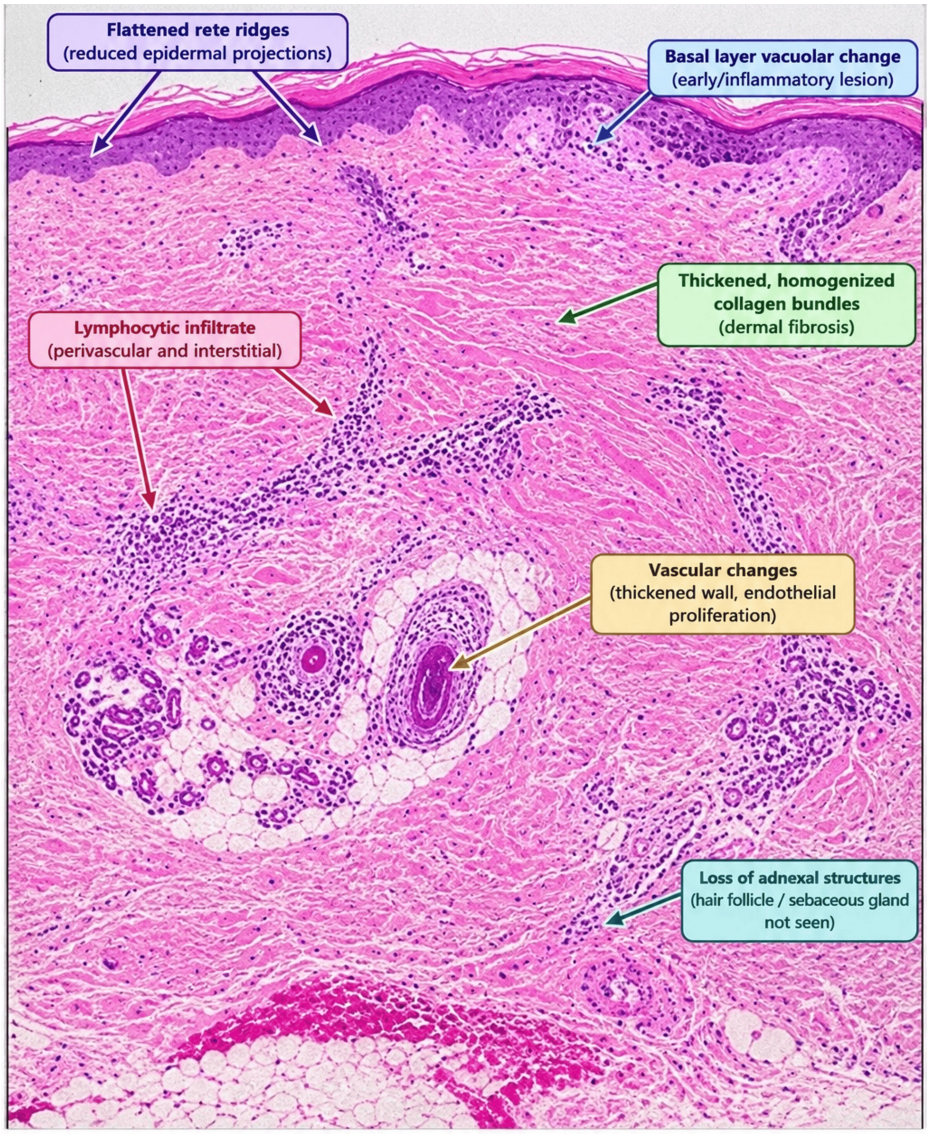


FIGURE 4: Hematoxylin and eosin (x100) stained skin biopsy showing flattening of rete ridges, basal layer vacuolar change, dermal fibrosis, and lymphocyte infiltrates consistent with systemic sclerosis

Differential diagnosis

Given the cutaneous stigmata of scleroderma, GAVE was the leading preendoscopic differential diagnosis. Peptic ulceration and reflux esophagitis were considered, both being frequent in SSc as a consequence of gastroesophageal reflux. Because of the splenomegaly, esophageal and gastric varices were regarded as close differentials. Telangiectasia-associated bleeding was also kept in mind, supported by the mucocutaneous findings and serology. Gastric or esophageal malignancy was considered less likely in the absence of weight loss, cachexia, or alarm symptoms, but could not be excluded without endoscopy. “A Mallory-Weiss tear” was thought unlikely given the absence of preceding retching. Table *1* lists the differential diagnoses and their resolution.

Differential diagnosis	Features in favor	Features against	Outcome
Gastric antral vascular ectasia (watermelon stomach)	Cutaneous features of systemic sclerosis; anemia; the commonest reported cause of gastrointestinal bleeding in this population	None on clinical grounds	Excluded at endoscopy
Esophageal or gastric varices	Splenomegaly; thrombocytopenia; hypoalbuminemia; sonographic chronic liver disease	No history of alcohol consumption or liver disease	Confirmed - bleeding gastric varices
Mucosal or submucosal telangiectasia	Mucocutaneous findings and serology of limited cutaneous systemic sclerosis	No telangiectatic lesions seen at endoscopy	Excluded
Peptic ulcer or erosive esophagitis	Gastroesophageal reflux is near universal in systemic sclerosis	No usage of nonsteroidal anti-inflammatory drugs; no ulcer or erosion at endoscopy	Excluded
Gastric or esophageal malignancy	Age; anemia	No weight loss, anorexia, cachexia, or dysphagia; no mass at endoscopy	Excluded
Mallory-Weiss tear	Hematemesis	No preceding retching or nonbloody vomiting; no mucosal tear at the gastroesophageal junction	Excluded

TABLE 1: Differential diagnosis of hematemesis in the present patient and its resolution

Management and outcome

Initial management comprised airway protection, intravenous fluid resuscitation, and acid suppression with an intravenous proton pump inhibitor. One unit of packed red blood cells was transfused in view of ongoing bleeding and tachycardia. Her blood pressure and heart rate stabilized, and urgent esophagogastroduodenoscopy was done within six hours of presentation (after hemodynamic stabilization), which revealed small esophageal varices and large gastric varices with active bleeding (Figure *5*); 1 mL of cyanoacrylate glue was injected, and hemostasis was achieved. No features of GAVE were seen (Figure *6*). Postprocedure, her vitals normalized, and hemoglobin showed a stable trend at 7.5 g/dL, then at 8 g/dL.

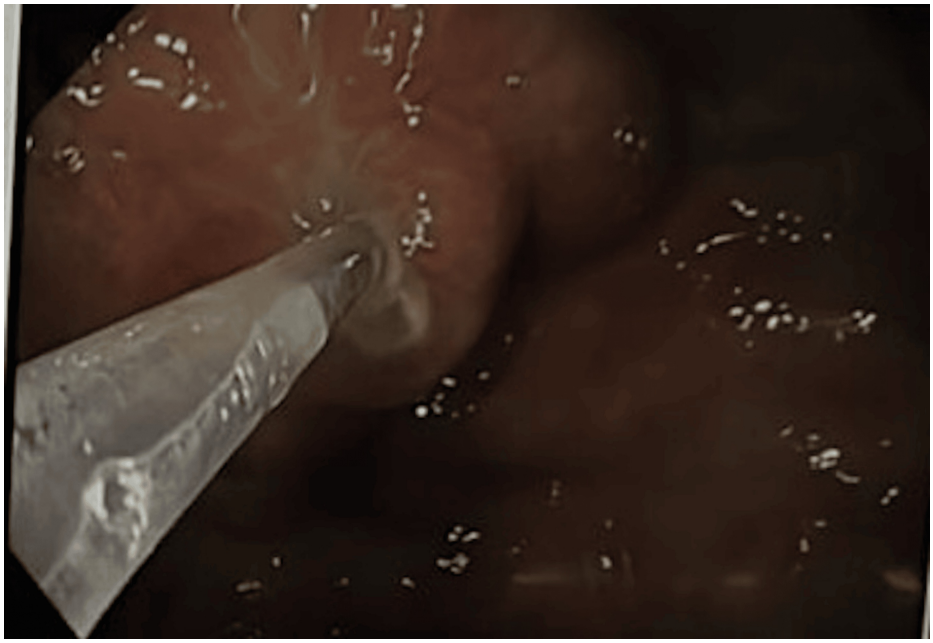


FIGURE 5: Esophagogastroduodenoscopy demonstrating large gastric varices (IGV-1 with signs of recent hemorrhage) with active bleeding, with glue being injected into the varix with a 21-gauge needle. No gastric antral vascular ectasia was identified

IGV-1: isolated gastric varix type 1

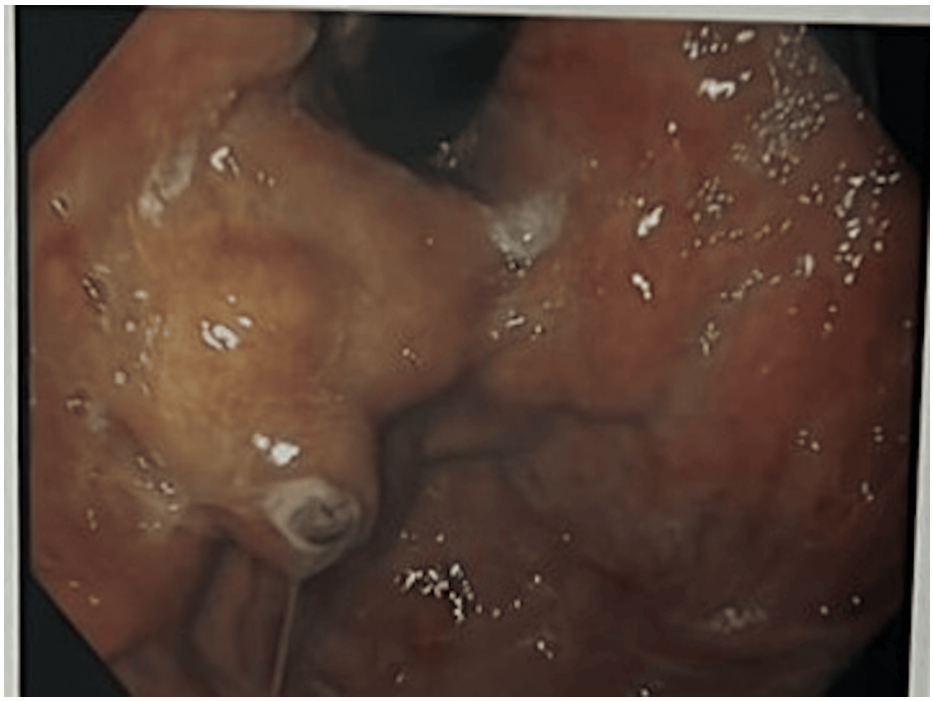


FIGURE 6: Esophagogastroduodenoscopy demonstrating large gastric varices (IGV-1 of size approximately 2 cm with signs of recent hemorrhage such as white nipple sign) (post-glue injection)

IGV-1: isolated gastric varix type 1

Given the massive gastrorenal shunt seen on computed tomography, PARTO was performed on day 5 as secondary prophylaxis. The gastrorenal shunt was obliterated by deployment of an Amplatzer vascular plug (12/9; Abbott Medical, Plymouth, MN) and sclerotherapy of gastric varices using Gelfoam (Pfizer Inc., New York, NY).

The patient was discharged on day 7 after stabilization on a nonselective beta-blocker, a proton pump inhibitor, hydroxychloroquine, and oral iron supplementation. She was reviewed two weeks after discharge, at which point clinical reassessment showed no recurrence of hematemesis and a stable hemoglobin level of 8 g/dL. A second review four weeks later confirmed continued improvement, with no further bleeding episodes and a hemoglobin level of 8.5 g/dL. The complete clinical course is summarized in Figure 7.

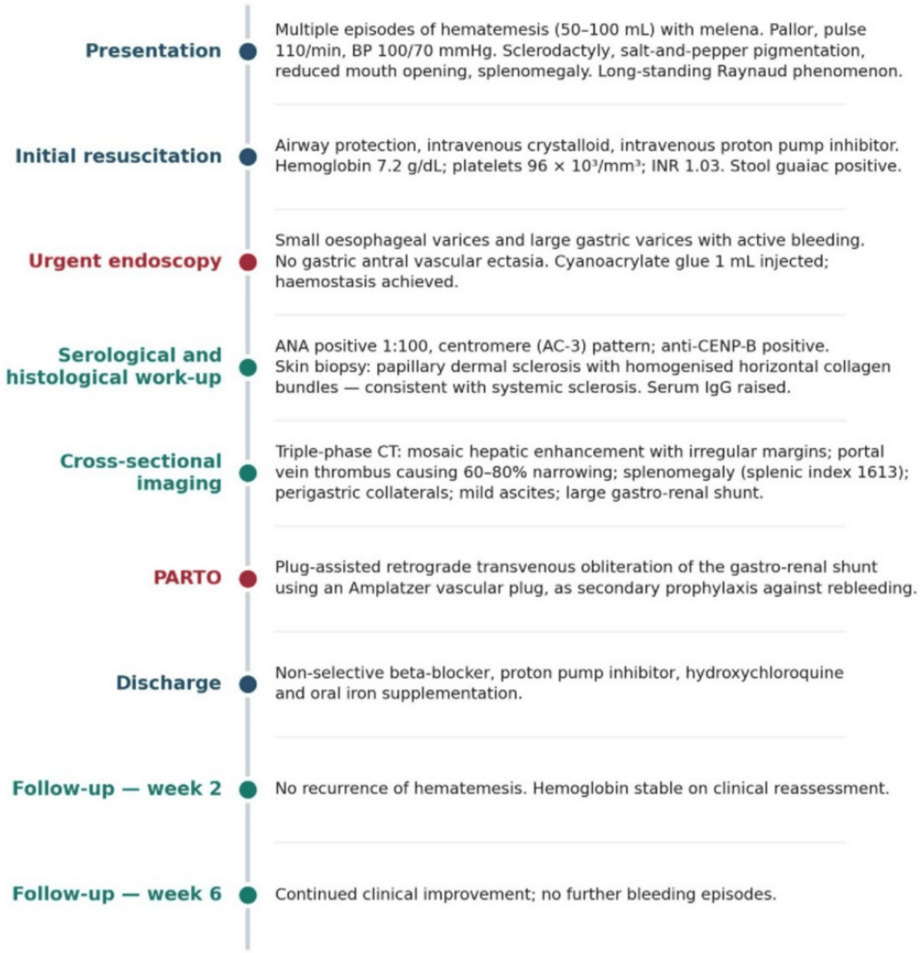


FIGURE 7: Chronological summary of the clinical course, from presentation with hematemesis through endoscopic hemostasis and endovascular shunt obliteration to six-week follow-up

PARTO: plug-assisted retrograde transvenous obliteration; ANA: antinuclear antibody; CENP-B: centromere protein B; BP: blood pressure; INR: international normalized ratio; IgG: immunoglobulin G; CT: computed tomography

Discussion

Gastrointestinal involvement in SSc is near universal and results from a sequence of vascular injury, neural dysfunction, smooth muscle atrophy, and fibrosis, which together impair motility and compromise mucosal integrity [11,12]. When overt bleeding occurs, the published literature has consistently directed attention toward scleroderma-specific mucosal lesions. Duchini and Sessoms attributed gastrointestinal hemorrhage in CREST syndrome principally to mucosal telangiectasia [5]; Miller et al. observed that gastrointestinal bleeding may be the first manifestation of SSc, particularly when telangiectatic lesions are submucosal [4]; and Nassar et al. described the increasing recognition of gastrointestinal bleeding in this population, linking it to progressive vascular damage [3]. Gastrointestinal disease is not a trivial complication: it is an independent contributor to morbidity and mortality in SSc [13].

The present case departs from that expectation in an instructive way. GAVE was our leading preendoscopic differential and was specifically excluded at endoscopy; the bleeding source was instead large gastric varices accompanied by small esophageal varices. The mechanism was therefore portal hypertensive rather than vasculopathic. With hindsight, the clues favoring portal hypertension were all present before endoscopy: thrombocytopenia disproportionate to the volume of blood loss, moderate splenomegaly, hypoalbuminemia, and sonographic evidence of chronic liver disease. These findings are easy to discount in a patient with SSc because anemia is readily ascribed to chronic occult blood loss from GAVE and thrombocytopenia to marrow suppression or immune destruction. We would argue that in any patient with SSc presenting with gastrointestinal hemorrhage, portal hypertension should be actively excluded before a scleroderma-specific lesion is assumed. Figure *8* outlines a suggested approach to a case of upper gastrointestinal bleeding in SSc.

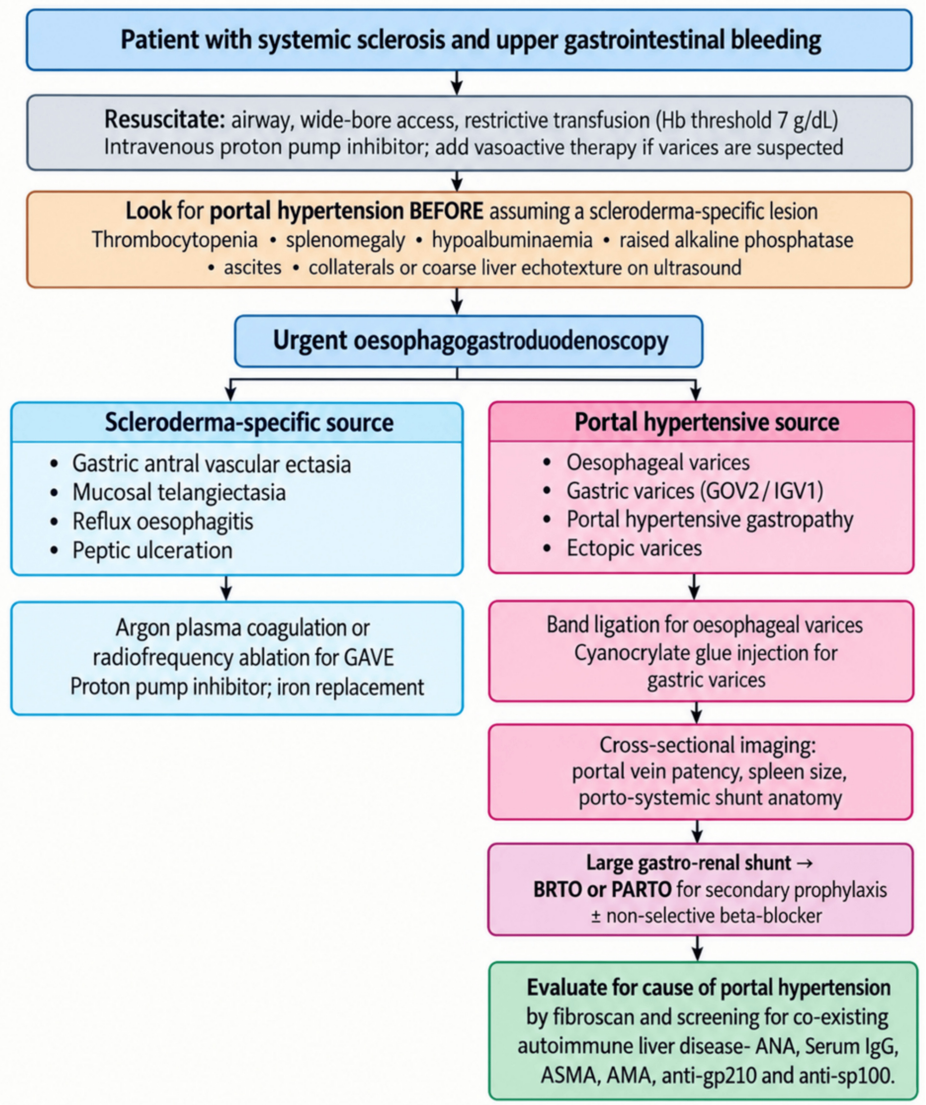


FIGURE 8: Proposed diagnostic and therapeutic approach to upper gastrointestinal bleeding in a patient with systemic sclerosis, emphasizing the assessment for portal hypertension before a scleroderma-specific mucosal lesion is assumed.

GAVE: gastric antral vascular ectasia; GOV2: gastroesophageal varix type 2; IGV1: isolated gastric varix type 1; BRTO: balloon-occluded retrograde transvenous obliteration; PARTO: plug-assisted retrograde transvenous obliteration; TIPS: transjugular intrahepatic portosystemic shunt; Hb: hemoglobin; ANA: antinuclear antibody; IgG: immunoglobulin G; ASMA: antismooth muscle antibody; AMA antimitochondrial antibody

The nature of the underlying liver disease merits scrutiny and is, in our view, the most important unresolved question raised by this case. The etiology of liver disease remained uncertain, although a diagnosis of autoimmune hepatitis-related chronic liver disease was inferred from the raised serum immunoglobulin G. However, the biochemical pattern was cholestatic rather than hepatitic: the alkaline phosphatase was

elevated at 209 IU/L while both transaminases and bilirubin were normal, and the total globulin fraction lay within the reference range. Anticentromere antibody positivity has a strong and long-established association with primary biliary cholangitis, and the coexistence of primary biliary cholangitis with limited cutaneous SSc constitutes Reynolds syndrome [8,9]. Primary biliary cholangitis-related autoantibodies are found in a considerable proportion of unselected SSc cohorts [10]. Antimitochondrial antibody, anti-gp210, and anti-sp100 were not tested in our patient, and in retrospect should have been, since a diagnosis of primary biliary cholangitis would have carried the specific therapeutic implication of ursodeoxycholic acid therapy. We report this openly as an instructive limitation: in a patient with anticentromere antibodies and a cholestatic liver profile, primary biliary cholangitis should be excluded before autoimmune hepatitis is accepted.

Portal vein thrombosis compounded the picture. Thrombosis of the portal vein complicates cirrhosis in a substantial minority of patients, driven by reduced portal flow velocity, endothelial injury, and the rebalanced hemostasis of chronic liver disease [14]. In our patient, the thrombus produced 60%-80% luminal narrowing of the main portal vein and near-complete occlusion of the right branch, which would have significantly augmented portal pressure and would have contributed to the development of the large gastrorenal shunt and enlargement of gastric varices that ultimately bled. Although the endothelial dysfunction intrinsic to SSc contributing to thrombus formation is speculative, it is biologically plausible given that microvasculopathy is a defining feature of the disease. As the patient does not have any previous radiological investigation record, it is assumed that liver cirrhosis led to the development of a portal vein thrombus, which augmented the portal pressure. Anticoagulation was not given as it was assumed to be a chronic portal vein thrombus with no superior mesenteric vein extension and no pain or mesenteric ischemia.

Gastric varices bleed less frequently than esophageal varices but do so more severely, with greater transfusion requirements and higher mortality [15]. Endoscopic injection of cyanoacrylate glue is the recommended first-line hemostatic therapy for actively bleeding gastric varices and was effective in our patient [16]. Rebleeding rates after glue injection alone nevertheless remain appreciable, which is why secondary prophylaxis was pursued. Where a large gastrorenal shunt is present (as it is in the majority of individuals with gastric varices), retrograde transvenous obliteration offers a durable solution. PARTO substitutes a vascular plug and gelatin sponge for the occlusion balloon and sclerosant used in conventional balloon-occluded retrograde transvenous obliteration, shortening the procedure, removing the need for a prolonged indwelling balloon, and avoiding sclerosant-related complications, with high reported technical success [17]. An important caveat is that obliterating a spontaneous portosystemic shunt raises portal pressure and may aggravate esophageal varices and ascites. As a result, our patient was released on a nonselective beta-blocker and requires ongoing endoscopic surveillance.

Adjunctive measures also warrant comment. Proton pump inhibitor therapy remains appropriate given that esophageal mucosal involvement is present in a large proportion of patients with SSc even while receiving acid suppression [18], and iron replacement addresses the cumulative deficit from occult and overt loss. Risk-stratification instruments such as the Rockall and Glasgow-Blatchford scores are useful adjuncts in the initial triage of upper gastrointestinal bleeding [19]. Hydroxychloroquine was also prescribed; while it is widely used for the musculoskeletal and overlap manifestations of connective tissue disease, randomized evidence supporting its use in SSc specifically is lacking, and the indication in an individual patient should be stated explicitly. Broader management of the gastrointestinal manifestations of SSc rests on prokinetic therapy, nutritional support, and treatment of small intestinal bacterial overgrowth [20,21], and where an obscure source is suspected after negative upper and lower endoscopy, capsule endoscopy has an established role [22].

Finally, this case underlines the value of multidisciplinary care. The diagnosis required dermatological and rheumatological input to characterize the connective tissue disease, gastroenterological expertise to secure hemostasis, and interventional radiology to deliver definitive secondary prophylaxis. Structured prediction and primary prevention of vascular complications are increasingly emphasized in the management of SSc [23], and a comparable structured approach to the assessment of portal hypertension in this population would be worthwhile.

Limitations

This is a single case report, and no causal inference can be drawn. Antinuclear antibodies specific to primary biliary cholangitis and antimitochondrial antibodies were not measured, so the precise nature of the underlying autoimmune liver disease remains unconfirmed, and liver biopsy was not performed. A thrombophilia evaluation for the portal vein thrombosis was not undertaken. Follow-up was extended to six weeks only, which is insufficient to assess the durability of the shunt obliteration or the long-term risk of aggravated esophageal varices.

Conclusions

Hematemesis in a patient with sclerodactyly should not be assumed to arise from GAVE or mucosal telangiectasia alone. This case demonstrates that limited cutaneous SSc may coexist with portal vein

thrombosis, portal hypertension, and chronic liver disease, producing gastric variceal hemorrhage that is managed by an entirely different diagnostic and therapeutic pathway. Straightforward bedside and laboratory clues, thrombocytopenia, splenomegaly, hypoalbuminemia, and a cholestatic liver profile should prompt evaluation for portal hypertension and for coexisting autoimmune liver disease, in particular primary biliary cholangitis, given its strong association with anticentromere antibodies. Urgent endoscopy establishes the source and secures hemostasis, and when there is a significant gastrorenal shunt, PARTO offers effective secondary prophylaxis. Screening endoscopy for residual varices can be done at regular intervals. Multidisciplinary management involving rheumatology, dermatology, gastroenterology, and interventional radiology in our case helped in better decision-making for achieving a favorable outcome.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Nidhesh Khemchandani

Acquisition, analysis, or interpretation of data: Nidhesh Khemchandani

Drafting of the manuscript: Nidhesh Khemchandani

Critical review of the manuscript for important intellectual content: Nidhesh Khemchandani

Supervision: Nidhesh Khemchandani

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References

1. van den Hoogen F, Khanna D, Fransen J, et al.: 2013 classification criteria for systemic sclerosis: an American College of Rheumatology/European League against Rheumatism collaborative initiative. Arthritis Rheum. 2013, 65:2737-47. 10.1002/art.38098
2. Fritzler MJ, Kinsella TD: The CREST syndrome: a distinct serologic entity with anticentromere antibodies . Am J Med. 1980, 69:520-6. 10.1016/0002-9343(80)90462-3
3. Nassar M, Ghernautan V, Nso N, et al.: Gastrointestinal involvement in systemic sclerosis: an updated review. Medicine (Baltimore). 2022, 101:e31780. 10.1097/MD.0000000000031780
4. Miller JB, Gandhi N, Clarke J, McMahan Z: Gastrointestinal involvement in systemic sclerosis: an update . J Clin Rheumatol. 2018, 24:328-37. 10.1097/RHU.0000000000000626
5. Duchini A, Sessoms SL: Gastrointestinal hemorrhage in patients with systemic sclerosis and CREST syndrome. Am J Gastroenterol. 1998, 93:1453-6. 10.1111/j.1572-0241.1998.00462.x
6. Ebert EC: Gastric and enteric involvement in progressive systemic sclerosis . J Clin Gastroenterol. 2008, 42:5-12. 10.1097/MCG.0b013e318042d625
7. Ingraham KM, O'Brien MS, Shenin M, Derk CT, Steen VD: Gastric antral vascular ectasia in systemic sclerosis: demographics and disease predictors. J Rheumatol. 2010, 37:603-7. 10.3899/jrheum.090600
8. Reynolds TB, Denison EK, Frankl HD, Lieberman FL, Peters RL: Primary biliary cirrhosis with scleroderma, Raynaud's phenomenon and telangiectasia. New syndrome. Am J Med. 1971, 50:302-12. 10.1016/0002-9343(71)90218-x
9. Rigamonti C, Bogdanos DP, Mytilinaiou MG, Smyk DS, Rigopoulou EI, Burroughs AK: Primary biliary cirrhosis associated with systemic sclerosis: diagnostic and clinical challenges. Int J Rheumatol. 2011, 2011:976427. 10.1155/2011/976427
10. Cavazzana I, Ceribelli A, Taraborelli M, et al.: Primary biliary cirrhosis-related autoantibodies in a large cohort of Italian patients with systemic sclerosis. J Rheumatol. 2011, 38:2180-5. 10.3899/jrheum.110167
11. Gyger G, Baron M: Gastrointestinal manifestations of scleroderma: recent progress in evaluation, pathogenesis, and management. Curr Rheumatol Rep. 2012, 14:22-9. 10.1007/s11926-011-0217-3
12. Marie I, Dominique S, Levesque H, Ducrotté P, Denis P, Hellot MF, Courtois H: Esophageal involvement and pulmonary manifestations in systemic sclerosis. Arthritis Rheum. 2001, 45:4-346.
13. Hoffmann-Vold AM, Volkmann ER: Gastrointestinal involvement in systemic sclerosis: effects on morbidity and mortality and new therapeutic approaches. J Scleroderma Relat Disord. 2021, 6:37-43. 10.1177/2397198319891282
14. Intagliata NM, Caldwell SH, Tripodi A: Diagnosis, development, and treatment of portal vein thrombosis in

patients with and without cirrhosis. Gastroenterology. 2019, 156:1582-99.e1. 10.1053/j.gastro.2019.01.265
15. Sarin SK, Lahoti D, Saxena SP, Murthy NS, Makwana UK: Prevalence, classification and natural history of gastric varices: a long-term follow-up study in 568 portal hypertension patients. Hepatology. 1992, 16:1343-9. 10.1002/hep.1840160607
16. Garcia-Tsao G, Abraldes JG, Berzigotti A, Bosch J: Portal hypertensive bleeding in cirrhosis: risk stratification, diagnosis, and management: 2016 practice guidance by the American Association for the Study of Liver Diseases. Hepatology. 2017, 65:310-35. 10.1002/hep.28906
17. Gwon DI, Kim YH, Ko GY, et al.: Vascular plug-assisted retrograde transvenous obliteration for the treatment of gastric varices and hepatic encephalopathy: a prospective multicenter study. J Vasc Interv Radiol. 2015, 26:1589-95. 10.1016/j.jvir.2015.07.011
18. Marie I, Ducrotte P, Denis P, Hellot MF, Levesque H: Oesophageal mucosal involvement in patients with systemic sclerosis receiving proton pump inhibitor therapy. Aliment Pharmacol Ther. 2006, 24:1593-601. 10.1111/j.1365-2036.2006.03180.x
19. Laine L, Jensen DM: Management of patients with ulcer bleeding . Am J Gastroenterol. 2012, 107:345-60; quiz 361. 10.1038/ajg.2011.480
20. Ghaith N, Sophia L, Chris TD: Management of gastrointestinal manifestations in systemic sclerosis (scleroderma). Int J Clin Rheumatol. 2013, 8:661-73.
21. Nadler M, Eliakim R: The role of capsule endoscopy in acute gastrointestinal bleeding . Ther Adv Gastroenterol. 2014, 7:87-92. 10.1177/1756283X13504727
22. Volkmann ER, McMahan Z: Gastrointestinal involvement in systemic sclerosis: pathogenesis, assessment and treatment. Curr Opin Rheumatol. 2022, 34:328-36. 10.1097/BOR.0000000000000899
23. Bruni C, Cometi L, Gigante A, Rosato E, Matucci-Cerinic M: Prediction and primary prevention of major vascular complications in systemic sclerosis. Eur J Intern Med. 2021, 87:51-8. 10.1016/j.ejim.2021.01.022