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Case Report

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Complexity Of Palliative Care And Need For Early Referral In Scleroderma

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To The Editor

Systemic sclerosis (SSc) is a rare, life-limiting connective tissue disease with high mortality and diverse manifestations. Despite significant symptom burden and an unpredictable course, palliative care remains underutilized in SSc, leaving many patients with unmet needs in symptom management, quality of life, and advance care planning **[1]**. Additionally, skin and gastrointestinal involvement may alter drug pharmacokinetics, complicating treatment. We present a case to underscore the critical role of specialist palliative care in managing the complex and evolving needs of SSc patients. Consent was given by patient's next of kin.

Abstract

Background

Patients with systemic sclerosis (SSc) are at high risk of sudden clinical deterioration, particularly in the context of multiorgan involvement. There is under-recognition of decline and a corresponding lack of timely referral to specialist palliative care services. This case highlights the complexities of symptom management in SSc, which are compounded by the underlying disease process and its impact on pharmacokinetics.

Case Presentation

Mrs. LP, a 73-year-old woman with multiple comorbidities, presented with her first episode of SSc. Her disease manifestations included pulmonary hypertension, Raynaud's phenomenon with digital ulceration, sclerodactyly, and gastrointestinal involvement. She required a prolonged hospital admission for management of SSc and its complications, which were further compounded by functional decline, psychological distress, and challenges in analgesia. Palliative care was consulted to assist with analgesia due to escalating pain despite high-dose oral opioids. Her pain was significantly better managed with subcutaneous morphine, likely due to impaired gastrointestinal absorption. Initially, Mrs. LP was reluctant to acknowledge her functional decline, reflecting difficulty in understanding the life-limiting nature of her illness. Through ongoing education, she developed greater insight into her condition and engaged in advanced care planning, which was formally documented. This proved invaluable during a subsequent episode of acute deterioration, guiding the consensus decision to transition to end-of-life care.

Conclusion

This case highlights the unique complexities of providing palliative care to patients with SSc. Such patients often present with multifaceted needs that require early recognition of clinical deterioration and timely referral to specialist palliative care services to optimise symptom management and support holistic care.

Keywords: scleroderma, palliative, pain, analgesia

Introduction

Patients with systemic sclerosis (SSc) are at high risk of sudden clinical deterioration, particularly in the context of multiorgan involvement. There is under-recognition of decline and a corresponding lack of timely referral to specialist palliative care services. This case highlights the complexities of symptom management in SSc, which are compounded by the underlying disease process and its impact on pharmacokinetics.

Case Description

A 73-year-old woman was admitted to our rheumatology unit with a new diagnosis of systemic sclerosis (SSc). Her complications included digital ulcers with osteomyelitis requiring amputation and IV antibiotics, sclerodactyly, and pulmonary hypertension managed with IV diuretics. During admission, she experienced a suspected esophageal variceal bleed; however, gastroscopy was deferred due to high anaesthetic risk. Her manifestations and associated investigations are listed in **Table 1**.

Table 1: summary of patient SSc manifestations

SSc manifestations	Severity and complications
Sclerodactyly	Modified Rodnan's skin score (mRSS) 18
Raynaud's phenomenon	Digital ulceration and osteomyelitis
Gastrointestinal	Vomiting, nausea, constipation, suspected malabsorption
Pulmonary hypertension	Pulmonary pressure of 66mmHg on transthoracic echocardiogram
Cardiac	Diastolic dysfunction transthoracic echocardiogram and pulmonary oedema on chest imaging requiring intravenous diuresis and high flow oxygen
Serology	ANA 1:640 centromere

She sustained right-sided rib fractures (5th, 10th, and 11th) after a ward fall and developed localized upper back and chest pain, described as dull with intermittent sharp episodes. Initial analgesia included Tapentadol SR 50 mg regularly, PRN oxycodone 5 mg hourly, and Tapentadol IR 50–100 mg every four hours, but her pain remained poorly controlled despite being opioid naive. PRN buprenorphine (200–400 mcg every three hours) was subsequently added. She was previously opioid-naïve.

Palliative care was consulted due to escalating pain, requiring 180 mg oral morphine equivalent daily. On assessment, the patient reported nausea, regurgitation, and constipation—likely related to SSc-related gastrointestinal involvement, previously overlooked amid repeated fasting for surgical planning. A subcutaneous syringe driver with morphine (initially 30 mg, titrated to 40 mg) provided effective pain relief, and regular laxatives were introduced.

Advance care planning discussions were initiated, which the patient welcomed in light of her acute decline and multi-organ involvement. She was later transferred to her rural local hospital, where she likely suffered a cardiac event. Thanks to early conversations with palliative care, she was promptly transitioned to end-of-life care and died peacefully, surrounded by loved ones. The family later expressed their appreciation of these early and realistic discussions.

Discussion

Palliative care aims to enhance quality of life by relieving suffering and supporting early advance care planning for patients with serious illnesses and their caregivers. Traditionally introduced when disease-modifying treatments ceased to be effective, its role is now evolving. A more contemporary model integrates palliative care alongside active treatment, recognizing that both approaches can be complementary. Patient care is not always linear—active management and palliative support can and should occur in parallel **[2]**.

Palliative care services are underutilised in the management of rheumatic conditions including systemic sclerosis (SSc). In a review of palliative care input in systemic rheumatic conditions **[1]** found that only 15% of patients received care from a palliative physician, most during their terminal admission, with a median referral just 8 days before death. Of those referred, 72% were only introduced to palliative care during their terminal admission, with a median referral time of 8 days before death. Despite this, many exhibited clear palliative needs—such as pain, dyspnoea, anorexia, and functional decline—at least a year prior. Notably, only 3.5% had completed advance care planning before their final admission.

Patients with SSc have significant unmet needs for specialist palliative care **[3]**. Common symptoms include fatigue, breathlessness, and constipation, with pulmonary, myositis, and gastrointestinal involvement being key predictors of palliative care requirements. The complexity and overlap of these symptoms highlight the importance of a multidisciplinary, SSc-specific palliative care approach. Early integration of palliative care at diagnosis, with ongoing assessment, can help monitor disease trajectory while addressing both symptom burden and disease-directed treatment **[4]**.

Several factors contribute to the underutilization of palliative care in SSc **[5]**. Disease progression is difficult to quantify, and clinicians often assess severity by individual organ systems, lacking a unified grading system to guide referral. The disease course is highly unpredictable, with extended periods between flares, making it challenging to identify when palliative care would be most beneficial **[6]**. Additionally, mortality in rheumatic diseases is often attributed to complications of immunosuppression or organ-specific issues, rather than the underlying disease, limiting recognition of end-of-life needs **[7]**. We recommend early introduction of palliative care for symptomatic support, especially during episodes of recurrent medical destabilisation and hospitalisation or failure of adequate disease control despite escalation of immunosuppression.

Current disease activity indices in SSc—such as the European Scleroderma Study Group (EScSG) index **[8]**, the 12-point Disease Activity Index (DAI) **[9]**, and the Combined Response Index for SSc (CRISS) **[10]**—focus on organ-specific involvement but do not capture symptom severity or the impact of multiorgan failure. While high activity correlates with disease burden, these tools often underestimate overall symptom complexity and psychological distress, potentially delaying palliative care referral **[11]**. In contrast, the Palliative Care Outcome Collaboration (PCOC) framework **[12]**, which tracks transitions from stable to unstable states, may better reflect symptom burden and guide timely palliative care involvement.

Symptom management in SSc is further complicated by GI dysmotility in SSc which significantly contributes to morbidity and mortality. Presentations are heterogeneous and may include gastroesophageal reflux, delayed gastric emptying, dysphagia, vomiting, diarrhea, malabsorption, and pseudo-obstruction. In our patient, this was initially unrecognised. GI dysfunction in SSc is thought to result from vascular atrophy, immune infiltration, smooth muscle atrophy, and neurofibrosis **[13]**. These changes impair peristalsis and digestion, leading to malabsorption and obstruction risk **[14]**. Oral medication absorption is suspected to be compromised—though this remains unquantified in current literature. Opiates are commonly used in palliative care for dyspnoea and pain. But their side effects—particularly delayed intestinal transit and constipation—are especially problematic in SSc patients. Due to impaired GI motility, opioids can worsen existing complications like constipation and pseudo-obstruction **[15]**. Moreover, there is limited evidence supporting opioid use for chronic pain in SSc, and their use carries a heightened risk of adverse events **[16]**.

Opiate titration was challenging in our case, reflecting common difficulties in SSc patients. GI complications can significantly alter the pharmacokinetics of oral medications, necessitating alternative administration routes and careful dose adjustments. Transdermal formulations may bypass GI issues and reduce related side effects, but their absorption can still be affected by scleroderma skin changes, fever, or sweating **[17]**.

In addition to GI absorption issues, SSc patients also have impaired sublingual absorption due to progressive vascular damage. This includes endothelial activation, apoptosis, intimal thickening, and reduced sublingual blood flow, as shown by laser Doppler studies **[18]**. They also exhibit decreased sublingual capillary density and thinner glycocalyx layers compared to healthy controls. Longer disease duration increases the risk of microhaemorrhages and capillary rarefaction.

We acknowledge that this is a case report written in retrospect following expressed appreciation towards the palliative care team from the patient's family members. Much of the data is retrospective and requires interpretation of clinical notes which may be subject to bias. We feel that a gentle introduction to supportive care via a palliative care physician in the outpatient setting would be more accepting to most patients.

Conclusion

In conclusion, early recognition of decline and timely specialist palliative care involvement are crucial in SSc management. We feel that this aspect of SSc management is underappreciated and requires increased resource allocation for both research and clinical management.

List Of Abbreviations

CRISS: Combined Response Index for SSc

DAI: Disease Activity Index

EScSG: European Scleroderma Study Group

GI: Gastrointestinal

mRSS: Modified Rodnan's skin score

PCOC: Palliative Care Outcome Collaboration

SSc: systemic sclerosis

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