CASE REPORT

The Emergence of Pott's Disease in Complicated Systemic Sclerosis

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Abstract

Systemic sclerosis (SSc) is a rare, autoimmune connective tissue disease that causes fibrosis and collagen deposition in the skin, joints, and internal viscera. Such patients are more prone to infections, which can affect almost any organ system. However, tuberculosis in SSc patients has rarely been reported. Involvement of the spine is rare in patients with TB and has never been priorly reported in a patient with SSc. A 42-year-old female with complicated SSc had interstitial lung disease, pulmonary hypertension, and Sjogren's syndrome; and presented with the complaint of backache and fever. Upon lab and radiological investigations, we elucidated a collapsed lumbar vertebra. She was subsequently diagnosed with Pott's disease. After conservative treatment and initiation of anti-tuberculous medications, her symptoms improved and she was discharged. We emphasize the significance of careful evaluation, radiological assessments, and TB screening in patients with autoimmune illnesses, especially in regions where TB is endemic.

Keywords: *Spinal TB, pott's disease, systemic sclerosis, tuberculosis.*

INTRODUCTION

Systemic sclerosis (SSc) is a multisystem, autoimmune disease with a global prevalence of around only 17-23 per 100,000 people [1, 2]. It is characterized by aberrant immune system activation which causes systemic fibrosis in almost all organs throughout the body [3].

SSc predisposes individuals to several infections: from uncomplicated urinary tract infections to severe sepsis [4]. However, the presence of tuberculosis (TB) in individuals with SSc has scarcely been described in prior literature [5]. TB, although primarily a pulmonary disease, is itself a multi-system pathology that can affect virtually any organ system in the body. However, extra-pulmonary manifestations are seen in only 3% of cases [6]. Even rarer, is spinal TB, also known as "Pott's disease", named after Sir Percivall Pott (1713-1788) who was a British surgeon, and accounts for 10% of extra-pulmonary manifestations in individuals with TB [6]. In this case report, we present a case of Pott's disease in a patient with SSc. To the best of our knowledge, such an association has not been previously described in medical literature.

CASE PRESENTATION

History and Examination

A 42-year-old female, who was previously diagnosed with systemic sclerosis (SSc) twelve years ago and interstitial lung disease (ILD) five years ago, presented to our outpatient department with fever and backache for the past 5 months. In addition, she had breathing difficulty with bilateral pedal swelling for the past 1 month. Her fever was low-grade and intermittent. She had also lost weight over the past months. Her backache was progressive and severe enough to limit her normal daily routine tasks. Initially, she had trouble breathing upon exertion, but recently she developed it at rest too. Her systemic history consisted of the following: orthopnea, paroxysmal nocturnal dyspnea, generalized skin tightening and thickening, joint pain (multiple), dysphagia, constipation, positive Raynaud's phenomenon, oral and ocular dryness, alopecia and photosensitivity.

Upon presentation, she was hypertensive (blood pressure, 160/100 mm Hg), tachypnic (respiratory rate, 22 per minute), and febrile (101 F). She was mildly anemic, had a raised jugular venous pressure, bilateral pitting pedal edema, restricted oral opening with a beaked nose, deformed distal interphalangeal joints with contractures of both hands (**Fig. 1**), generalized tight and thick skin, hypo- and hyper-pigmented scars with self-amputation of 2nd digit of the left, and 3rd digit of right foot (**Fig. 1**). On musculoskeletal testing, there were overall restricted joint movements because of skin thickening, and severe tenderness at the back. Respiratory examination revealed reduced air entry bilaterally, with fine crepitations in the lower zones of both lungs. There was a loud P2 audible. Her abdominal examination was unremarkable.

Diagnostics and Work-up

Based on careful evaluation, an initial impression of SSc with pulmonary and cardiac involvement was made. Subsequently, lab investigations were ordered. Her labs showed microcytic, hypochromic anemia with

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Received: June 26, 2024; Revised: December 05, 2024; Accepted: December 09, 2024
DOI: https://doi.org/10.37184/jlnh.2959-1805.3.3

  ISSN: 2960-2963 (Online) *Journal of Liaquat National Hospital 2025; 3(2): 152-156*

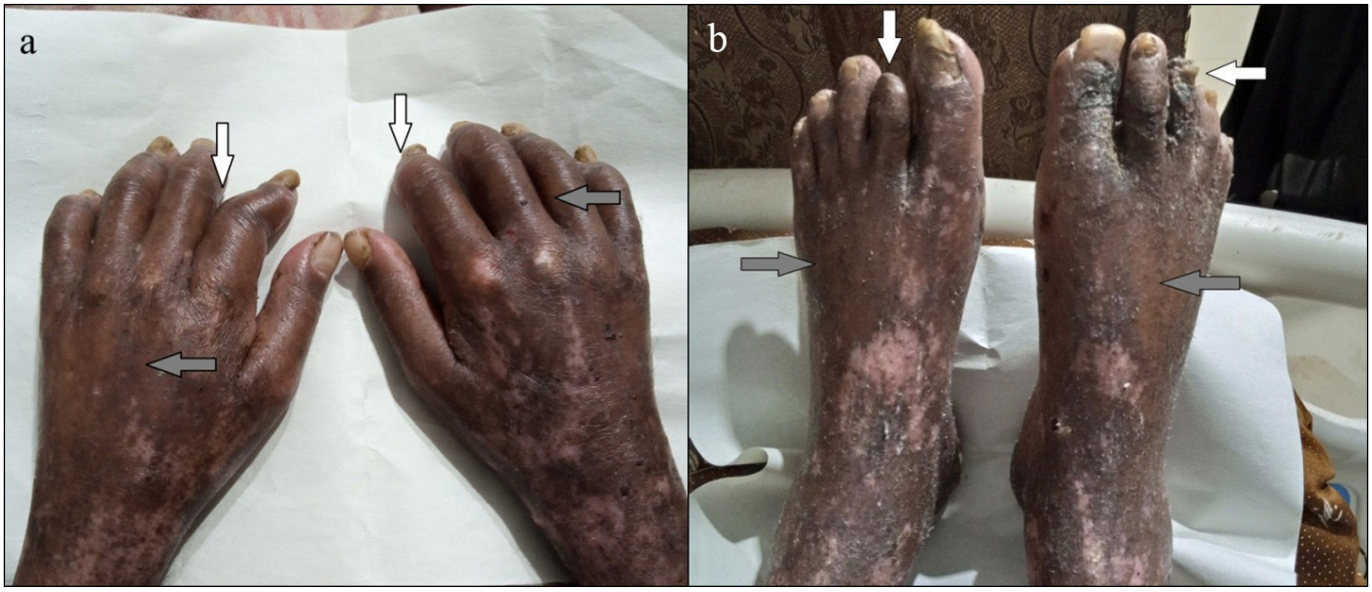

Fig. (1): Manifestations of typical systemic sclerosis features in the extremities: **(a)** Deformed distal interphalangeal joints (white arrows) of the second digits, with contractures, in both hands. Skin tightening, visible as loss of wrinkles, along with a shiny appearance can also be appreciated on the dorsum of the left hand, and all digits (grey arrows); **(b)** Auto-amputation of the 2nd digit of the left foot (white arrow), and the 3rd digit of the right foot (white arrow) can be seen. In addition, skin tightening (grey arrows) can be appreciated on the dorsum of both feet.

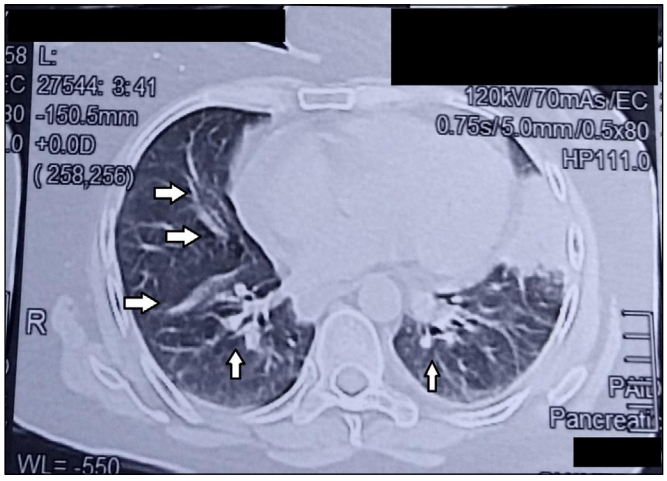

Fig. (2): High-resolution computed tomography (HRCT) showing bilateral lower lobes with fibrotic changes (multiple white arrows).

a haemoglobin of 8.8 g/dL, with normal white blood cell and platelet counts. Her remaining baselines were unremarkable, but her CRP was 75, and her ESR was 110. Her connective tissue disorder workup revealed: antinuclear antibody, positive; anti-dsDNA, 2.3 IU/mL (normal, <20 IU/mL); U1-RNP antibody, 0.8 (normal, <3.2 IU/mL); SS-A, 15.5 (normal, <3.2 IU/mL); SS-B, 18.6 (normal, <8 IU/mL); Scl-70 antibodies, 67 IU/mL (normal, <3.2 IU/mL); anti-CCP, normal; RA factor, normal.

Upon radiological workup, her high-resolution computed tomography (**Fig. 2**) showed patchy areas of ground glass haze in bilateral lower lobes with fibrotic changes, and the findings were consistent with ILD. Her echocardiography revealed left ventricular dysfunction, mild pericardial effusion, pulmonary arterial systolic pressure of 65 mmHg, and an ejection fraction of 25%. Magnetic resonance imaging (MRI) showed lumbar spine evinced collapse of the L3 vertebral body (**Fig. 3**) and showed bilateral collections which were noted more on the left psoas muscle. The MRI showed transverse processes with a narrow neural foramen suggestive of Pott's disease. Interestingly, sputum, and gastric lavage, for acid-fast bacilli and GeneXpert were negative. However, after consultation with radiology, other pathologies like osteoporotic fractures *etc.,* were ruled out and a diagnosis of Pott's disease was made.

Treatment and Clinical Course

Based on the workup, we diagnosed our patient with complicated SSc with Sjogren's syndrome, ILD, pulmonary hypertension, with concurrent Pott's disease. She was conservatively managed with anti-TB therapy (pyrazinamide 400mg, ethambutol 275mg, rifampicin 150mg, and isoniazid 75mg) for Pott's disease, capsule omeprazole 40mg OD before breakfast, tablet domperidone 10mg BD, and syrup lactulose 30ml HS for dysphagia and constipation, tablet ramipril 10mg OD for hypertension, tablet nifedipine 30mg OD for ischemic digital ulcers, tablet bosentan 62.5mg BD for raised pulmonary artery pressure, and analgesics for joint pain. Anti-TB medicines were prescribed as three tablets before breakfast. For ILD, the tablet prednisolone at 5mg OD and tablet azathiaprone at 50 mg OD were continued as per the previous regimen. She experienced an improvement in her symptoms after a few days of

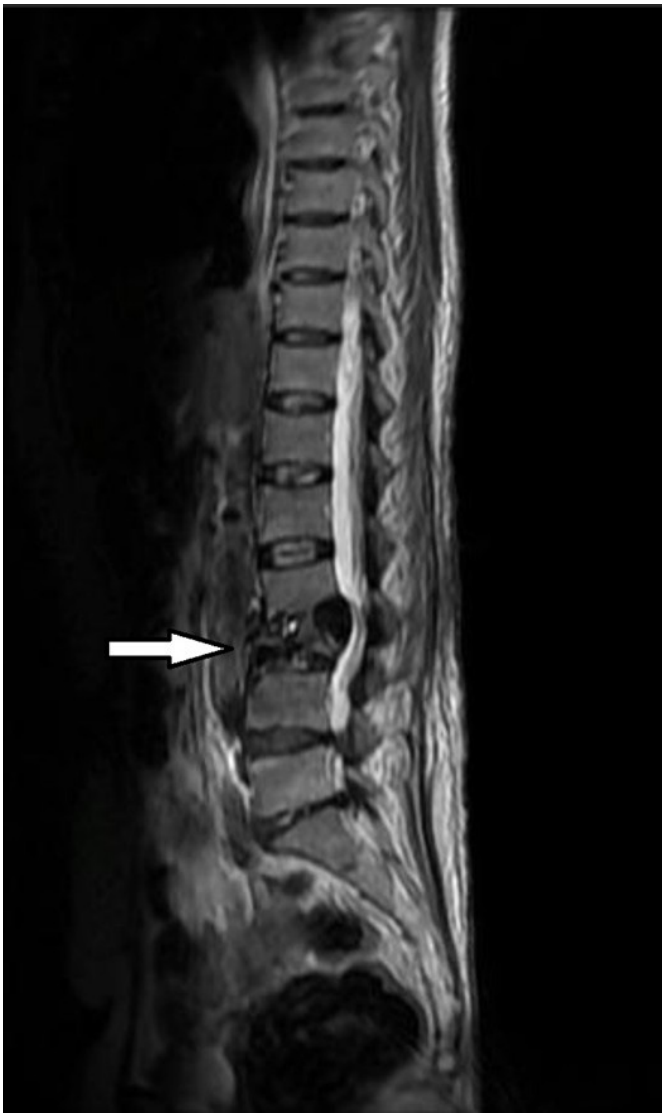

Fig. (3): Sagittal view of the spine upon magnetic resonance imaging (MRI) which shows the collapse of the L3 vertebral body (white arrow).

treatment and was subsequently discharged. Now, she is on regular follow-up *via* the outpatient department.

DISCUSSION

SSc alongside tuberculosis (TB) has rarely been documented in medical literature. Published literature consists of case reports [7], case series [8], and a retrospective study [5, 9]. However, there is no prior reported study of SSc alongside vertebral TB. The relevance and importance of our case report is not limited to its novelty, but also to the challenging nature of the case, as it describes a complicated case of SSc where several organ systems were involved, and was also, additionally, coupled to Pott's disease.

Pott's disease is named after Sir Percivall Pott who first described it in 1779 [10]. This rare manifestation of extra-pulmonary TB presents with nonspecific symptoms which are chiefly limited to back pain and fever [10]. Our case presented similarly with backache and fever (see History and Examination), albeit other symptoms attributable to other comorbid diseases were also present. Additionally, radiographs have limited specificity which is why MRI is indicated to provide a more reliable assessment of invasion depth and surrounding tissue damage - which often spares the disk [10]. It should also be noted that an absence of the causative organism in microbiological results should not exclude Pott's disease from the differential diagnoses [10]. The management of our case was in line with these methods.

Our patient had several features which are typical of SSc. Such a Raynaud phenomenon occurs in nearly all SSc patients. Patients may experience prolonged ischemic tissue loss, unlike those with primary (idiopathic) Raynaud's phenomenon. Our patient had joint contractures (**Fig. 1**) and a positive history of Raynaud's phenomenon on exposure to cold and in winter. In our case, the presentation was severe owing to auto-amputation of the toes (**Fig. 1**). SSc also affects the cardiac system, by causing cardiovascular fibrosis, myositis, anomalies in the conduction system, coronary artery disease, pericardial effusion, and cardiac failure [11]. Our patient had mild pericardial effusion and cardiac failure with a severely decreased ejection fraction of 25%. Furthermore, pulmonary involvement, such as ILD and raised pulmonary artery pressure are now the most common causes of death in SSc [12]. Consistent with established associations, our patient's HRCT (**Fig. 2**) showed ILD, alongside pulmonary hypertension with a pressure of 65 mmHg as evinced by echocardiography.

In SSc, the entire gastrointestinal tract is impacted. Patients experience gastroesophageal reflux, constipation due to a hypomotile gut, and bloating or diarrhea due to bacterial overgrowth. Our patient was constipated and had complaints of dysphagia and indigestion. Although SSc patients generally experience hypertensive crises, our patient only presented with a raised blood pressure which was managed with ramipril. Another manifestation of SSc is painful and stiff joints. In our case, acro-osteolysis, which refers to bone resorption, of the distal phalanx was evident.

It is well-established that SSc predisposes individuals to infections. This can be attributed either to the pathophysiology of the disease or to the use of immunosuppressant drugs which inadvertently encourage infections [13]. For example, cutaneous involvement, especially in the presence of calcinosis, makes an individual more prone to developing cellulitis,

and in rare cases, necrotizing fascitis [13]. Involvement of the gastrointestinal tract can manifest in the form of small intestinal bacterial overgrowth, Helicobacter Pylori infection, and/or rarely esophageal candidiasis [13]. Similarly, lung involvement as in the case of ILD, predisposes SSc patients to respiratory tract infections. This can manifest in the form of pneumonia, lung abscess, or empyema [13].

Pulmonary infections in SSc patients warrant special attention in the context of our case, but more importantly so because pneumonia is reportedly one of the most common severe infections in SSc patients [4]. Pneumonia is more common in patients who develop ILD, which could be due to structural alterations, in addition to the aberrant immune response. Esophageal dysmotility may also contribute to the pathophysiology of pneumonia owing to aspiration in SSc patients. A study of 117 SSc patients found a positive association between esophageal dysmotility and aspiration pneumonia (odds ratio, 1.23) [13].

In light of the above, infection with Mycobacterium tuberculosis has also been reported in medical literature. SSc patients are 2.53 times more likely to develop pulmonary TB [5]. Although there is not a significant association between SSc and extrapulmonary TB, patients with SSc are more likely to develop such manifestations [5]. A multivariate analysis in a study elucidated the following risk factors for TB infection in SSc patients: age greater than 60 years; presence of pulmonary arterial hypertension, and corticosteroid use [5]. In a recent Indian report, TB was the most commonly associated infection in SSc, and was primarily located in the lungs; although only 29.3% of SSc patients were affected [9]. The emergence of TB can be linked to the use of immunosuppressant drugs. In our case, our patient was already taking azathioprine and prednisolone for her ILD, which has been highlighted as a significant risk factor previously [5].

However, one must interpret the discussed information with caution, as studies that sought to evaluate TB infection in SSc patients are scarce. One might even question the role of TB in the pathophysiology and development of SSc, especially in countries where TB is endemic.

CONCLUSION

Environmental factors play a probable role in the development of SSc, and infections by various viruses and bacteria are also thought to contribute. Molecular mimicry, superantigens, and microchimerism are likely mechanisms due to which agents like human parvovirus B19, cytomegalovirus, Ebstien-barr virus, and Endogenous retrovirus are hailed as likely infectious etiologies in the development of SSc [14, 15]. Therefore, further research is needed to elucidate the exact relationship between TB and SSc, especially in consideration of the endemic nature of Mycobacterium tuberculosis.

CONSENT FOR PUBLICATION

Written informed consent was obtained by authors from the patients prior to submission of the case report.

CONFLICT OF INTEREST

The authors declare that they have no competing interests.

ACKNOWLEDGEMENTS

Declared none.

AUTHORS' CONTRIBUTION

Muhammad Luqman: contributed to case identification and manuscript writing.

Muhammad Abdul Rehman: was involved in drafting the manuscript and conducting a critical review.

Zasheer Fatima: was responsible for collecting clinical data.

Hafiz Muhammad Faizan Abid: participated in clinical data collection and assisted in manuscript drafting.

Syed Muhammad Kashif: provided a critical review of the manuscript.

Muhammad Tanveer Alam: critically evaluated the case and granted final approval for submission.

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