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CASE REPORT

First reported case of Erasmus Syndrome in Iran: [^{18}F]FDG PET/CT findings in silicosis-associated systemic sclerosis

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ABSTRACT

A 53-year-old man with a 20-year history of occupational silica exposure presented with progressive dyspnea, unexplained weight loss, skin thickening of the fingers, and Raynaud's phenomenon. Initial HRCT findings were consistent with fibrotic interstitial lung disease and calcified mediastinal lymph nodes related to silica exposure. Given the patient's unexplained weight loss, whole-body [^{18}F]FDG PET/CT was performed to exclude occult malignancy. The PET/CT scan demonstrated no evidence of a metabolically active primary tumor or metastatic disease; however, multiple metabolically active pulmonary nodules, fibroreticular parenchymal changes, and calcified mediastinal lymph nodes were identified. Based on the clinical and imaging findings, the patient fulfilled the 2013 ACR/EULAR classification criteria for systemic sclerosis. In the context of the patient's occupational history and clinical features, these findings supported the diagnosis of Erasmus syndrome. A brief review of the literature indicates that, to the best of our knowledge, no cases from Iran have been reported to date. This case underscores the added value of [^{18}F]FDG PET/CT in the differential diagnosis of pulmonary nodules and mediastinal lymphadenopathy, particularly in excluding malignancy and characterizing inflammatory and fibrotic disease activity, thereby facilitating more accurate diagnosis and appropriate therapeutic decision-making in complex occupational and autoimmune lung diseases.

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INTRODUCTION

Erasmus syndrome represents an uncommon clinical condition in which systemic scleroderma develops after occupational exposure to silica, first described in 1957 [1, 2]. Occupational exposure to silica is a well-established hazard in industries and linked to the onset of silicosis, a progressive fibrotic lung disorder [3]. Epidemiological evidence indicates that, in addition to pulmonary disease, inhalation of crystalline silica is strongly associated with a range of autoimmune disorders, particularly systemic sclerosis [4].

Scleroderma is an autoimmune disorder defined by immune system imbalance, skin thickening, and fibrosis of internal organs [2, 3]. Among the complications of systemic sclerosis, interstitial lung disease (ILD) is considered the most severe, responsible for approximately one-third of disease-related-deaths. Although high-resolution computed tomography (HRCT) remains the standard tool for diagnosing and monitoring ILD, [^{18}F]FDG PET/CT provides additional non-invasive insights into ILD severity and prognosis [5, 6].

To date, only a limited number of Erasmus syndrome cases have been reported, predominantly from South Asia [1–3]. Notably, these reports were based primarily on clinical, serologic, and HRCT findings. This report describes a patient with Erasmus syndrome following long-term occupational silica exposure. To the best of our knowledge, this is the first reported case from Iran which highlight the potential role of [^{18}F]FDG PET/CT in the comprehensive assessment and differential diagnosis of Erasmus syndrome.

CASE PRESENTATION

A 53-year-old-man with approximately two decades of occupational exposure to silica dust manifested with progressive dyspnea and unexplained weight loss. HRCT of the chest demonstrated fibrotic changes and parenchymal scarring in both upper lobes, particularly the apical and posterior segments. Also, multiple calcified lymph nodes are noted in the mediastinum and bilateral hila, consistent with pneumoconiosis (Figures 1 and 2).

The diagnosis of systemic sclerosis was established according to the 2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria [7]. The patient fulfilled the classification criteria with a cumulative score of 14 points, exceeding the threshold of ≥9 required for definite systemic sclerosis. The contributing items included skin thickening of the fingers extending proximal to the metacarpophalangeal joints (9 points), Raynaud's phenomenon (3 points), and LID confirmed by HRCT (2 points). A structured summary of the fulfilled ACR/EULAR criteria is provided in Table 1. Serologic testing demonstrated a strongly positive antinuclear antibody (ANA) titer (>1:1600), which further supported the diagnosis of systemic sclerosis. Though, anti-Scl-70 antibody was detected at a borderline level (17.4 U/mL), this finding did not contribute additional points to the classification score.

According to unexplained weight loss, whole-body [^{18}F]FDG PET/CT was performed to exclude underlying malignancy. The patient fasted for 6 hours prior to the scan, and the blood glucose level was 85 mg/dL before tracer injection. After intravenous administration of 8.5 mCi (0.1 mCi/kg) of [^{18}F]FDG, the patient rested in a quiet room during the uptake phase, and imaging was initiated 60 minutes after injection. Whole-body PET/CT acquisition was performed using a Siemens Biograph 6 TruePoint scanner.

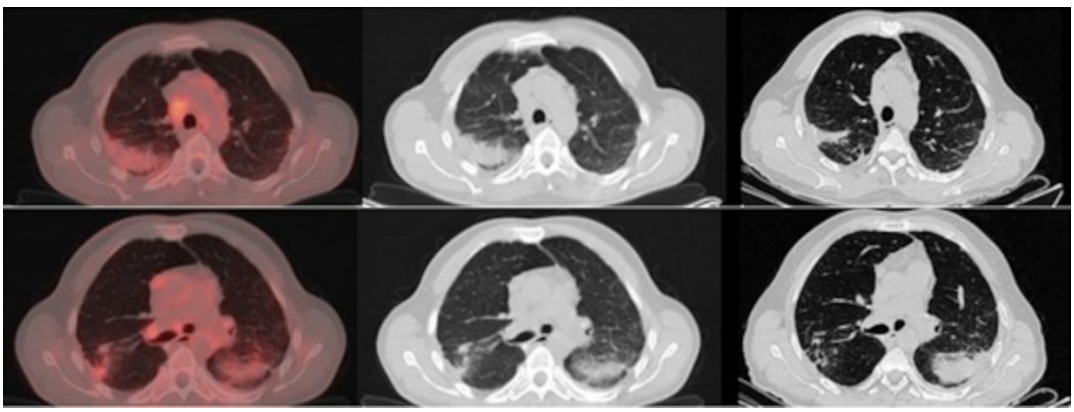

Figure 1. [^{18}F]FDG PET/CT and HRCT images evaluation of pulmonary parenchymal changes. From left to right: axial fused [^{18}F]FDG PET/CT, low-dose CT, and corresponding HRCT images demonstrate peripherally located metabolically active fibroreticular consolidations in the bilateral lungs

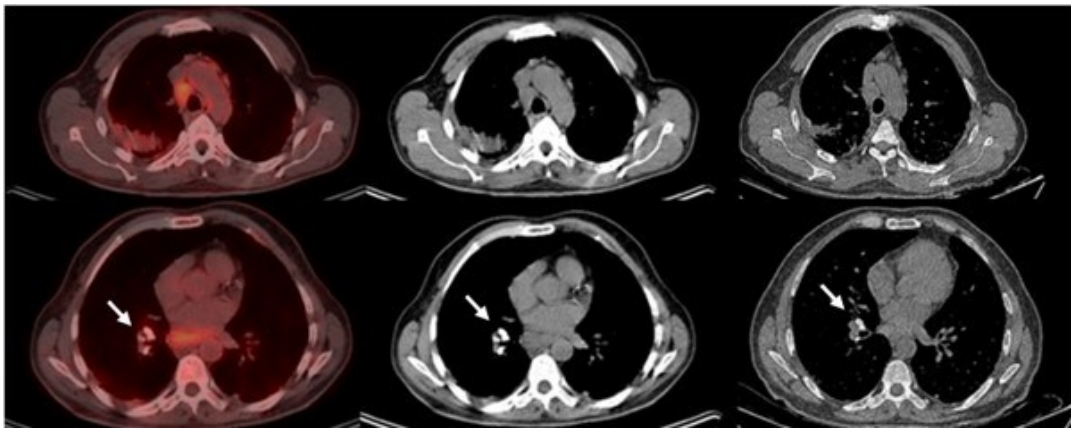

Figure 2. [^{18}F]FDG PET/CT and HRCT evaluation of mediastinal lymphadenopathy. From left to right: axial fused [^{18}F]FDG PET/CT, low-dose CT, and corresponding HRCT images. The PET/CT fusion images reveal moderate metabolic activity within the mediastinal lymph nodes, which also demonstrate characteristic "eggshell" calcifications identifiable on both PET/CT and HRCT scans (arrows)

PET data were acquired in 3D high-definition mode with an acquisition time of 3 minutes per bed position. A low-dose non-contrast CT scan (30 mAs, 110 kV, 5-mm slice thickness, pitch 1.2) was obtained for attenuation correction and anatomical localization. PET images were reconstructed using an iterative reconstruction algorithm (2 iterations, 21 subsets), Gaussian filter (FWHM = 5) and a 168 × 168 matrix without time-of-flight capability. Resolution recovery was applied using TrueX.

The [^{18}F]FDG PET/CT scan revealed no clear evidence of malignancy; however, it demonstrated multiple pulmonary micronodules with minimal metabolically activity in both lungs, predominantly in the mid and lower lobes. Peripheral consolidative changes with mild metabolic activity were noted in the posterior segment of the right upper lobe (SUVmax = 2.4) and in the apicoposterior segment of the left upper lobe adjacent to the oblique fissure (SUVmax = 2.3), accompanied by peripheral fibrotic changes, subpleural reticulation, and mild interlobular septal thickening (Figure 1). Also, multiple mediastinal and hilar lymph nodes with variable size and [^{18}F]FDG uptake were identified, several of which showed dense calcification showing "eggshell" pattern in the prevascular, paraaortic, subaortic, paratracheal, subcarinal, bilateral hilar/peribronchial, and paraesophageal regions. The most prominent findings were metabolically active matted lymph nodes in the right paratracheal region with SUVmax up to 4.2, measuring up to 27 × 17 mm, and in the subcarinal regions with SUVmax up to 3.8 (Figure 2). For reference, the hepatic background SUVmax was 2.2, and the mediastinal blood pool SUVmax was 1.8, confirming that pulmonary and nodal uptake was above physiologic background activity. Taken together, the clinical history of prolonged silica exposure, clinical features of systemic sclerosis, HRCT findings of ILD, and [^{18}F]FDG PET/CT demonstration of metabolically active pulmonary and nodal lesions supported the diagnosis of Erasmus syndrome, defined as the coexistence of systemic sclerosis and silicosis.

A literature search was performed in PubMed/MEDLINE, Scopus, Embase, and the Cochrane Library from database inception to December 2025 using combinations of the terms "Erasmus syndrome," "systemic sclerosis," "silica exposure," and "silicosis." To the best of our knowledge, this represents the first reported case of Erasmus syndrome from Iran and describes the corresponding [^{18}F]FDG PET/CT findings.

Inhalation of crystalline silica can trigger immune dysregulation through activation of macrophages, persistent inflammatory responses, and stimulation of fibroblast proliferation, ultimately contributing to the development of systemic sclerosis and lung fibrosis. Previous studies have suggested that patients with systemic sclerosis and occupational silica exposure may experience a more severe disease phenotype, frequently manifested with diffuse skin involvement and interstitial lung disease [8–10].

Although HRCT remains the cornerstone for the diagnosis and longitudinal monitoring of systemic sclerosis-associated interstitial lung disease (SSc-ILD), it provides primarily morphological data. Recent evidence indicates that [^{18}F]FDG PET/CT serves as a valuable functional adjunct, reflecting inflammatory and metabolic activity within fibrotic lung tissue, typically manifesting as increased uptake in the basal lung segments and mediastinal lymph nodes. This elevated [^{18}F]FDG uptake is closely linked to macrophage activation and active inflammatory processes. Furthermore, studies have demonstrated that [^{18}F]FDG PET/CT is a sensitive tool for assessing disease activity, aiding in patient

stratification, and potentially monitoring treatment response, particularly in patients with early or progressive SSc-ILD [11, 12].

Several alternative diagnoses were excluded based on clinical and imaging criteria. Sarcoidosis was considered because [18F]FDG-avid mediastinal and hilar lymphadenopathy is a typical feature of active granulomatous disease. However, although lymph node calcification can be seen in long-standing sarcoidosis, it usually shows irregular or amorphous patterns rather than classic eggshell morphology. In contrast, silica-related pneumoconiosis is presented with dense, often peripheral "eggshell" calcifications. The absence of systemic granulomatous manifestations (e.g., ocular, cutaneous, cardiac, or neurologic involvement) and the lack of a pattern suggestive of highly active granulomatous inflammation further argued against sarcoidosis [13].

Tuberculosis was also considered in the differential diagnosis, as [18F]FDG PET/CT can demonstrate [18F]FDG-avid lesions in both active and, less commonly, subclinical or early-reactivating latent disease. However, active pulmonary tuberculosis typically presents with characteristic imaging features such as cavitation, focal or multifocal consolidation, tree-in-bud opacities, or a miliary reticulonodular pattern, frequently accompanied by intensely [18F]FDG-avid lymphadenopathy. Conversely, chronic post-tuberculous sequelae manifest as fibrotic scarring, volume loss, and calcified parenchymal or nodal lesions—often with a characteristic upper-lobe predominant distribution—which typically show low or absent metabolic activity on PET/CT, reflecting inactive disease rather than ongoing inflammation [14]. Given the absence of either active or typical chronic TB patterns, combined with the patient's clear occupational history and autoimmune background, tuberculosis was considered an unlikely diagnosis [14].

Progressive massive fibrosis (PMF), representing complicated silicosis, was another important differential diagnosis. PMF is classically defined by large, often symmetrical, mass-like opacities in the bilateral lungs, frequently containing air bronchograms and dense internal calcifications [15]. In contrast, the imaging pattern in this case was not that of massive parenchymal fibrosis but rather with fibroreticular changes of interstitial lung disease compatible with systemic sclerosis (SSc-ILD) superimposed on silica-related damage, which is more consistent with Erasmus syndrome than isolated PMF.

Occupational lung cancers may mimic pneumoconiosis on CT, but they more commonly present with a dominant mass, asymmetric focal uptake, and higher [18F]FDG activity, and malignant nodal involvement rarely shows dense calcification [15]. The combination of only moderate metabolic activity, absence of a primary tumor or dominant mass, and densely calcified mediastinal lymph nodes favored a chronic occupational inflammatory process rather than a malignant etiology.

Taken together, when interpreted in the context of long-term silica exposure and established systemic sclerosis, the lack of defining features of sarcoidosis, tuberculosis, PMF, and malignancy supports Erasmus syndrome as the most plausible and unifying diagnosis.

Table 1. Application of the 2013 ACR/EULAR classification criteria for systemic sclerosis in our case. The patient fulfilled the classification requirements with a total score of 14, exceeding the threshold of ≥9 for a definite diagnosis of systemic sclerosis [7]

Criteria domain	Weight	Patient's findings	Allocated score
Skin thickening	9		9
Finger lesions	2		-
Telangiectasia	2		-
Abnormal nailfold capillaries	2		-
Lung involvement	2		2
Raynaud's phenomenon	3		3
SSc-related antibodies	3	Anti-Scl 70 = 17.4 U/ml (>18)	-
Total cumulative score	≥ 9		14

ACR/EULAR: American College of Rheumatology/European Alliance of Associations for Rheumatology; Anti-Scl 70: anti-topoisomerase I antibody; SSc: Systemic sclerosis

CONCLUSION

This case highlights the association between chronic occupational silica exposure and the development of systemic sclerosis, known as Erasmus syndrome. It underscores the importance of careful correlation between imaging findings and occupational history when evaluating patients with interstitial lung disease. It also demonstrates the complementary role of [^{18}F]FDG PET/CT in the differential diagnosis of pulmonary nodules and mediastinal lymphadenopathy, particularly in excluding malignancy and in assessing inflammatory and fibrotic activity to facilitate a more accurate diagnosis and appropriate clinical approach in complex occupational and autoimmune lung diseases.

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