Morphea Sculpted in Silica: A Case Report of Limited Cutaneous Systemic Sclerosis in a Woman with Long-Time Exposure to Silica Dust

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Localized cutaneous systemic sclerosis is a subset of systemic sclerosis that has several clinical subtypes, which can be a challenge to diagnose. An association of environmental and occupational determinants to systemic sclerosis has been widely suggested, and in a recent meta-analysis, it was strongly demonstrated. Although silica dust is often described as an occupational risk factor for diffuse cutaneous systemic sclerosis, few case reports have been published about it in relation to morphea. Here, we present a case of a sculptor with localized scleroderma. Sculptors are known to be at high risk for developing scleroderma, which is related to their constant contact with silica particles.

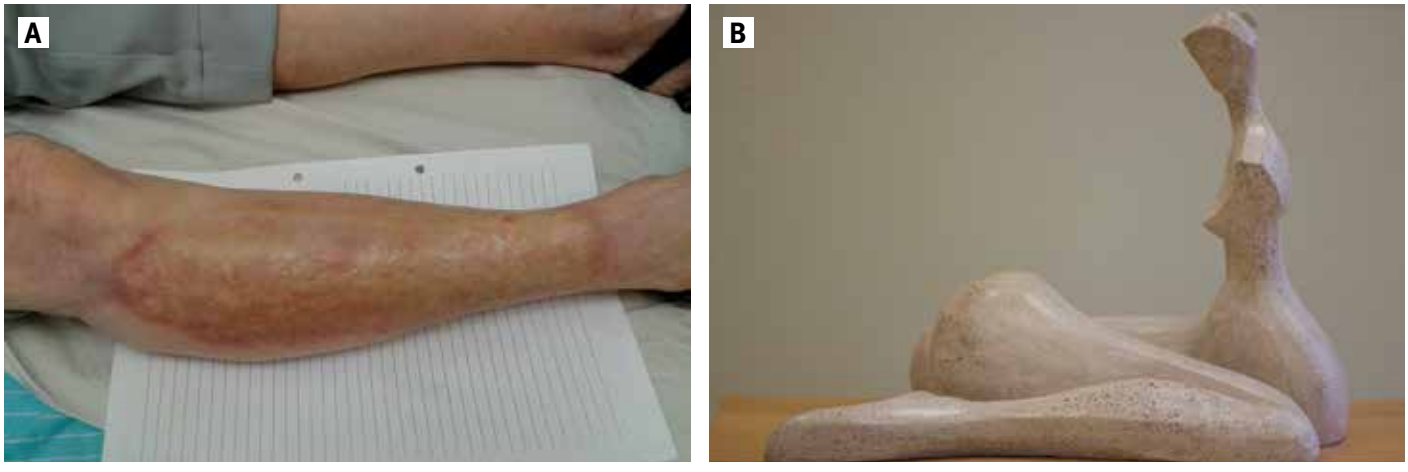
PATIENT DESCRIPTION

A 93 years old woman with hypertension had been diagnosed with morphea on her right leg, which she had endured for the past 20 years [Figure 1A]. The diagnosis was proven by a skin biopsy of the lesion. Eight years ago, the lesion expanded and the margins had become erythematous and warm to physical touch. All blood tests, even the serologic ones, were normal. A trial with rituximab was started, without clinical improvement. Twenty years ago, she started to work as a sculptor.

COMMENT

Systemic sclerosis (SSc) is a rare autoimmune connective tissue disease [1]. It is classified into two different clinical subsets depending on skin involvement [2]. Limited cutaneous SSc (lcSSc), or morphea, is a subset characterized by skin sclerosis of the hands and feet distal to the elbows and knees, respectively. It can also affect the face [2]. lcSSc is a poorly understood subgroup that has various clinical subtypes, which can be distinguished by its clinical manifestations and the structure of the skin and subcutaneous tissue afflicted by the fibrosis [2]. However, sometimes classifications can be difficult to determine because the boundaries between the subtypes are not always clear [2]. lcSSc is usually associated with anti-centromere and anti-Th/To

Figure 1. [A] Indurated skin on the right leg (photography by Itamar Goldstein, M.D.) **[B]** Sculpture made by the artist (photography by João Pedro Gomes, M.D.)



antibodies, and there is no effective and universal therapy [2].

The etiology of lcSSc remains unclear [2], but an association between SSc and silica dust has been widely reported and a recent meta-analysis demonstrated this phenomenon [3]. This meta-analysis included 19 studies with more than 1300 people diagnosed with SSc and five case-control studies with an overall odd-ratio of 2.81, 95% confidence interval (CI) 1.86–4.23, $P < 0.001$ [3]. The symptoms are indistinguishable from those with classic SSc, and the autoimmune profile is similar [3]. Different case reports have found a relation between the SSc subgroup and silica dust exposure [4]. Silica particles are smaller than 1 μm and are the more pathogenic than bigger ones. They can cause lung disease, but they can also penetrate the skin of exposed areas [5]. However, bigger particles (1–20 μm) have been isolated on the skin of the arms of patients with occupational scleroderma [5]. The phagocytosis of the particles by macrophages stimulates a cytokine release, especially interleukin (IL)-1 and IL-6, which stimulate fibroblasts collagen synthesis [2]. Upon the death of the macrophages, silica is released, which perpetuates a vicious cycle [4]. The risk of SSc appears to be strongly associated with high cumulative exposure, which reflects the fact that silica is toxic to macrophages in a dose-dependent way [5]. It has been reported that people working in several occupations have a high risk of developing SSc because of the intense contact with silica. One such occupation is that of a sculptor [3].

The appearance of lcSSc in sculptors has been reported before, and it has been postulated that there is a direct role between lcSSc and dispersed silica particles [4]. In this case report, we discuss an elderly woman with directed contact to silica dust, who was diagnosed with morphea. She did not have evidence of systemic affliction, but the reason she developed lcSSc remains uncertain. The physical characteristics of the particles and the complete composition of the clay used by the sculptor might be the reason for this clinical manifestation.

CONCLUSIONS

More studies are needed to determine the characteristics of silica or the genetic susceptibly to skin exposure of silica dust.

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Capsule

Reduction in spinal radiographic progression in ankylosing spondylitis patients receiving prolonged treatment with tumor necrosis factor inhibitors

To evaluate the course of spinal radiographic progression for up to 8 years of follow-up in a large cohort of ankylosing spondylitis (AS) patients treated with tumor necrosis factor (TNF) inhibitors, Maas et al. enrolled consecutive patients from the Groningen Leeuwarden AS cohort starting TNF inhibitors between 2004 and 2012 in the study. Baseline and biannual radiographs were randomized with radiographs of TNF-naive AS patients and scored in chronologic order according to modified Stoke Ankylosing Spondylitis Spine Score (mSASSS). The course of radiographic progression (linear or non-linear) was investigated using generalized estimating equations. Primary analysis was performed in patients with complete data over 4, 6, and 8 years of follow-up. Sensitivity analysis was performed after single linear imputation of missing radiographic data and after adjusting for patient characteristics with possible influence on radiographic progression. At baseline, median mSASSS of 210 included AS patients was 2.8 (interquartile range 0.0–12.0), mean ± standard deviation mSASSS 10.0 ± 15.5. During the first 4 years, radiographic progression followed a linear course (estimated mean progression rate was 1.7 for 0–2 and 2–4 years). A deflection from a linear course was found in patients with complete and imputed data over 6 and 8 years. The estimated mean 2 year progression rate reduced from 2.3 to 0.8 in patients with complete 8-year data. The same pattern was found after adjustment for baseline mSASSS scores, presence of syndesmophytes, gender, HLA–B27 status, age, symptom duration, smoking duration, body mass index, disease activity, and nonsteroidal antiinflammatory drug use.

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Eitan Israeli

"I am always ready to learn, but I do not always like being taught"

Winston Churchill (1874–1965), British statesman, Prime Minister of the United Kingdom, won the Nobel Prize in Literature in 1953 for his overall lifetime body of work