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Severe Alopecia Complicating Systemic Sclerosis

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Authors' contributions

This work was carried out and written in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aims: To describe a case of systemic sclerosis (SSc) associated with severe alopecia areata (AA) responsive to topical and systemic treatments, including vasoactive and immunosuppressive drugs (mycophenolate mofetil).
Presentation of the Case: A 56 year old woman, affected by SSc as from 5 years back, developed a rapid hair loss that progressively involved a large area of the scalp. AA was diagnosed, after the exclusion of an overlapping systemic lupus erythematosus or fungal infection. Treatment with topical steroids and minoxidil, plus mycophenolate mofetil that was introduced for interstitial lung disease, led to progressive improvement of alopecia up to a complete resolution within 4 months.
Discussion: This is an interesting observation of SSc complicated by severe AA, which is often observed in patients affected by various autoimmune disorders. A possible common pathogenesis of AA and SSc is also discussed.

Keywords: Alopecia; scleroderma; systemic sclerosis.

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1. INTRODUCTION

Systemic sclerosis (SSc) is a connective tissue disease characterized by collagen overproduction by altered fibroblasts and microvascular abnormalities, responsible for both skin and visceral organ involvement [1]. While typical cutaneous manifestations are largely described [1], data about the involvement of the scalp in scleroderma patients are scarce. Alopecia areata (AA) is a common, non-scarring dermatologic condition, characterized by patches of scalp hair loss; disease duration varies from self-resolving to permanent, while the distribution may be focal or widespread to total body. AA is often associated with psychological morbidity that may negatively affect the patient's quality of life [2,3]. AA is a heterogeneous disorder, characterized by chronic inflammation of the hair follicles, the etiology of which remains unclear, and the pathogenesis is probably multifactorial. The disease might be the consequence of immune-system alterations triggered by different genetic, environmental, infectious, and/or physical (trauma) co-factors [3]. Clinical observations revealed that patients with AA are often diagnosed together with one or more autoimmune disorders, including systemic lupus erythematosus (SLE), myasthenia gravis, ulcerative colitis, vitiligo, type 1 diabetes, thyroiditis, celiac disease, and rheumatoid arthritis [3,4]. The association of AA with localized scleroderma has also been reported, while no cases of AA are described in SSc, to our knowledge, up to date. Here, we describe a patient with SSc complicated by AA of the scalp.

2. PRESENTATION OF CASE

A 56 year old woman was firstly referred to the Rheumatology Unit of the University of Modena in 2010. Her past clinical history was characterized by Raynaud's phenomenon from 2009. She developed skin sclerosis of the hands, forearms and face, and telangiectasias; moreover, at the time of the diagnosis, typical scleroderma pattern at videocapillaroscopy (late pattern) and positivity to anti-Scl70 antibodies were found. After few years, interstitial lung disease was detected by high resolution computerised tomography. No significant heart involvement, including signs of pulmonary hypertension at echocardiography, was found during the follow-up. The patient was first treated with calcium channel blockers. During the following years, she suffered recurrent digital ulcers, thus iloprost and bosentan were added to treatment.

In January 2014, the patient rapidly developed hair loss, which progressively involved a large area of the scalp (Fig. 1a); moreover, a few erythematous scaly, flaky, macular lesions appeared on the neck, the trunk, and the arms. Thus, in the suspicion of a lupus-like cutaneous manifestation, a diagnostic skin biopsy was performed. Histopathological examination revealed a reduction of the hair follicle cells with inflammatory infiltrate in and around the bulbar region of hair follicle, along with diffuse collagen infiltration of the dermis, consistent with the underlying SSc; furthermore, few fungal hyphae were found. On the basis of these findings, the case was collegially discussed with the dermatologists, and diagnosis of AA complicating SSc was made, since the presence of hyphae was not sufficient to diagnose active cutaneous fungal infection.

Patient's serology was re-evaluated, excluding the presence of anti-dsDNA autoantibodies or complement consumption. Overall, the suspicion of an overlapping syndrome SLE-SSc was reasonably excluded.

Topical steroids and minoxidil were administered in the ensuing months. Moreover, given the coincident progression of patient's interstitial lung disease, mycophenolate mofetil (2 g bid) was also added to the therapy. In agreement with the dermatologists, no anti-fungal treatment was administered. A slow progressive improvement of alopecia was observed in the ensuing 4 months, leading to a complete resolution (Fig. 1c).

3. DISCUSSION

To the best of our knowledge this is the first observation of severe AA associated with SSc, even if several hair disorders, including AA, can be found in different autoimmune systemic diseases and localized scleroderma (3, 4, 7-9). Recent clinical investigations considered AA an autoimmune disorder due to T cell-mediated hair follicle damage. In particular Xing et al. demonstrated that cytotoxic CD8(+)/NKG2D(+) T cells were both necessary and sufficient for the induction of AA in mouse models of disease [5]. In our case, the chronic inflammation of hair follicles due to the pre-existent seborrhoeic dermatitis might be considered as the against hair follicles was born, then controlled

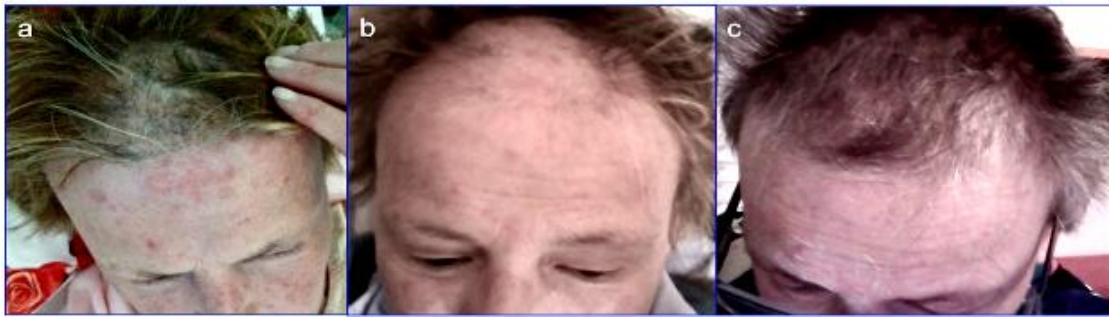

Fig. 1. Alopecia Areata in a patient with systemic sclerosis:
a. hair loss of the scalp
b. hair loss progressively involved the scalp; and
c. Significant improvement after combined therapy with topical corticosteroids and minoxidil, and systemic therapy with mycophenolate mofetil

with the local and systemic immunosuppressive therapy. Alternatively, a transient stressful condition as etiologic trigger could be hypothesized; nonetheless, a mere coincidental coexistence of AA and SSc cannot be excluded.

Alopecia was firstly described in patients with localized form of scleroderma in 1962 [6]; this possible association was also observed also in the linear subset of scleroderma "en coup de sabre" and morphea [7,8], two conditions that only sporadically may evolve to SSc [9]. We could speculate that both fibrotic and immune-mediated alterations of SSc might lead to severe injury and disappearance of hair follicles, even if exceptional, and with the possible contribution of unknown co-factors.

No clearly effective treatments for AA are available at the present time. Current management of AA is primarily symptomatic and often directed to mitigate the psychological distress [2]. In our patient the combined therapy with vasoactive and immunosuppressive drugs lead to improvement (Fig. 1c) of hair loss. Even if we can not exclude that alopecia could be improved spontaneously, combined therapy with minoxidil, mycophenolate mofetil, and iloprost might have contributed to this improvement. However, data from literature on the therapeutic effects of mycophenolate mofetil, iloprost, and other prostaglandin analogues in AA are conflicting [10,11].

4. CONCLUSION

This case might be regarded as a peculiar overlap of AA appearing in the context of SSc, possibly with the pathogenetic link of autoimmunity. Alternatively, SSc and AA may be just coincident. Clinicians should be aware of this rare possibility.

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

The authors have obtained the necessary ethical approval from the Institutional Committee. Moreover, this study is not against the public interest, and the release of information is allowed by legislation.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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