Deep necrotising morphea mimicking dermatitis artefacta

M. Melato, N. Gorji, O. Marangoni and G. Pillitteri

SUMMARY

The case of a 45-year old woman affected by facial ulcerative dermatitis diagnosed as dermatitis artefacta was reconsidered on the basis of the histological findings, which were consistent with a very unusual type of morphoae. The extraordinary features of this case, which we suggest calling »deep necrotising morphoea«, explain the diagnostic pitfall while highlighting the inadequacy of current classification of morphoea.

KEY WORDS

morphoea, dermatitis artefacta

Introduction

In 1954, in a basic paper on scleroderma, Leinwand et al. asserted that "In reviewing the history of scleroderma one is impressed by the methamorphosis of opinion regarding the classification of this disease" (1). 41 years later, other authors confirmed the persistency of an incomplete and confusing classification of morphoea and, while proposing a new one, they observed that the categories they were describing were not necessarily mutually exclusive and that the subtypes often co-occurred in the same patient (2). Therefore no exhaustive classification clearly distinguishes the different existing subtypes of morphoea, especially morphoea characterized by unusual features, exists at present and this fact may determine diagnostic pitfalls. There exist cases characterized by unusual features, which may present diagnostic pitfalls.

Case report

A 45-year-old woman noted the presence of a small facial dyschromia associated with deep induration, which in 4 months became complicated by blistering. Because of the progression of the lesion to a facial ulcerative dermatitis, a year later she was examined at a dermatological unit where the lesion was diagnosed as pathomimic ulcer of the face and treated with local antibiotic therapy. 11 months later, the patient was referred to one of the Authors (G.P.) complaining of progression of the disease complicated by facial sensitivity to cold and warm. The examination revealed in the right cheek a well demarcated, rather linear, subcutaneous induration covered by skin containing a superficial fistula discharging necrotic whitish material. The enlarging lesion showed a delicate violaceous border. Other symptoms and signs, including Raynaud's pheno-

menon, were lacking. Laboratory tests disclosed normal or negative CBC with differential count, urinalysis, liver function tests, rheumatic factor, levels of C3 and C4, and LE cell tests; antinuclear, anti-DNA, anti-Borrelia-burgdorferi, anti-toxoplasma IgG and IgM antibodies as well as anti-HIV antibodies were negative. ESR was elevated.

An adequate biopsy sample taken from the right cheek showed flattened epidermis. Dermis and septa of the subcutaneous fat were composed mainly of wavy collagen fibers, faintly stained with hematoxylin-eosin except at the borders, where there were more fibroblast and newly formed thick capillaries infiltrated by a sprinkling of lymphocytes and plasma cells. Dermal appendages were absent and there was no evidence of mucin (Fig. 1). Collagen bundles were seen deeply into the subcutaneous tissues where they encircled and separated the muscle fibres, causing atropia and necrosis (Fig. 2A & B); vessels and nerves were also involved by the fibrosis and deep arterioles showed hyperplastic walls (Fig. 3A & B). Homogenization of collagen bundles occurred especially in the deep dermis and subcutis. A necrotic area, poorly circumscribed by macrophages, epithelioid cells, and few eosinophils, was present superficially. Some black, finely granular, foreign material was intermingled with the necrotic tissue (Fig. 4), but no trace of it was found elsewhere.

When the woman was asked about the possible source of the foreign material, she hypothesized that it came from the toner cartridges from printers, as she had been working (exposure to solvents) as a cleaner in a bank for five years. She denied self-inflicted injury and use/abuse of colored chemicals or cosmetic substances. A month later the disease had progressed and two new ulcerative lesions had developed in the left face: the first was oval and located in the zygomatico-orbital region and the second, rather linear, in the naso-orbital region (Fig. 5). In the following twelve months the clinical features did not improve.

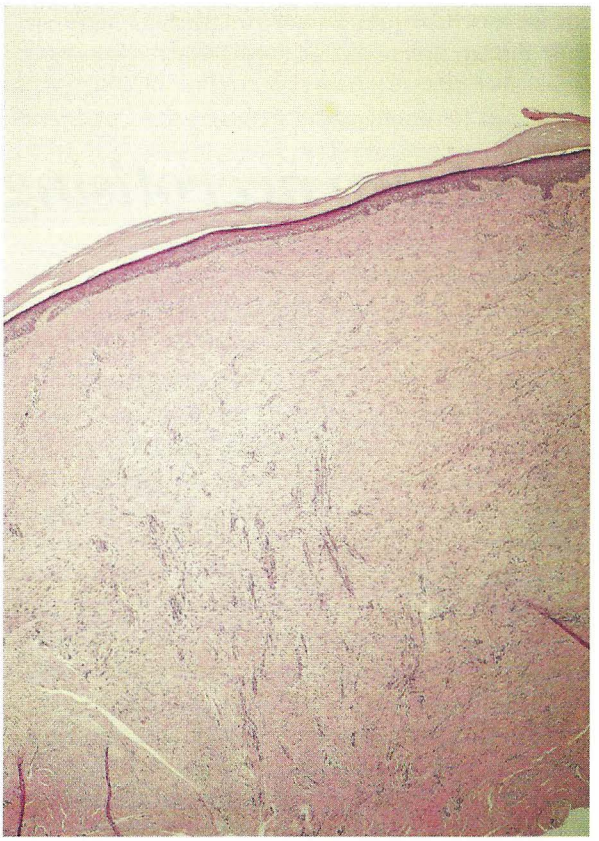
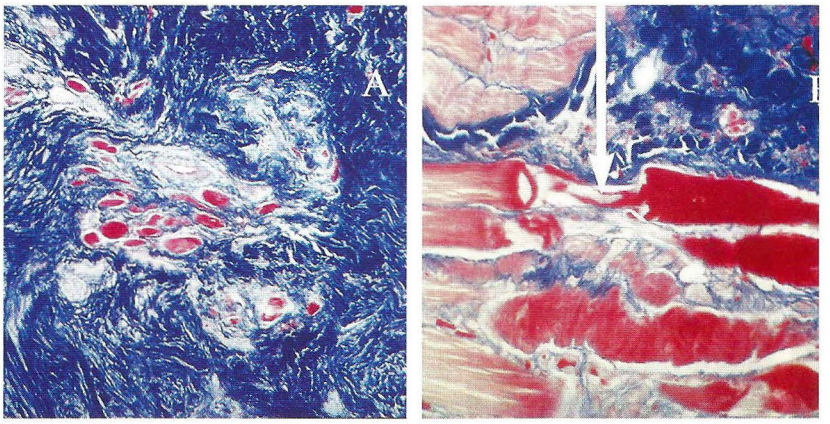


Figure 1. The epidermis is flattened; dermis and subcutaneous fat are replaced by collagen (H&E; original magnification X2.5).

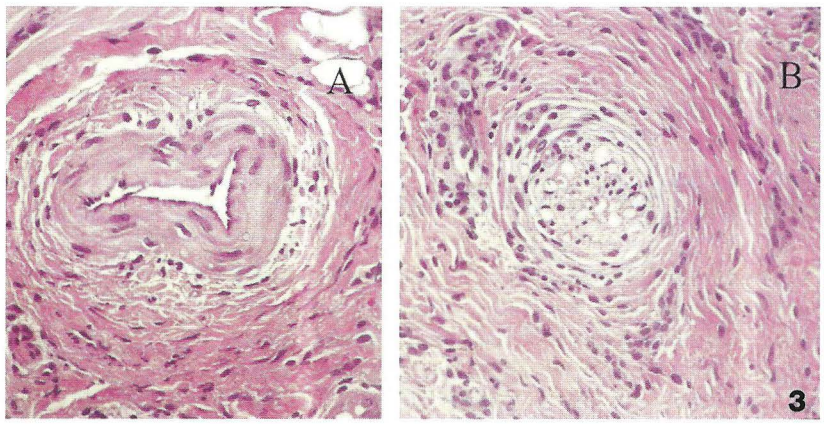
Figure 2. Collagen bundles penetrate deeply into the subcutaneous tissues where they encircle and separate the muscle fibers (A); in B, the arrow indicates where a muscle fiber is constricted by fibrous tissue and undergoes necrosis (AZAN-MALLORY; original magnification A X 2.5 and B X 40).

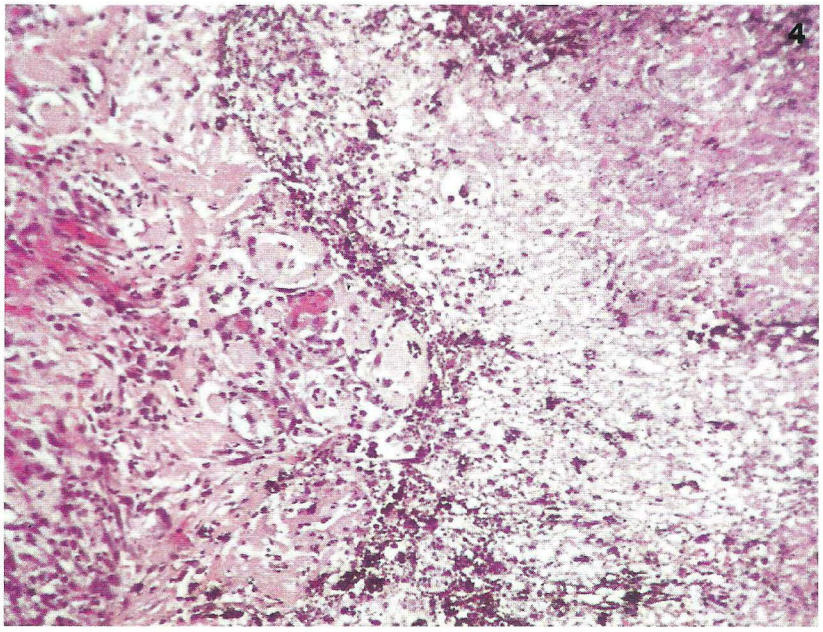


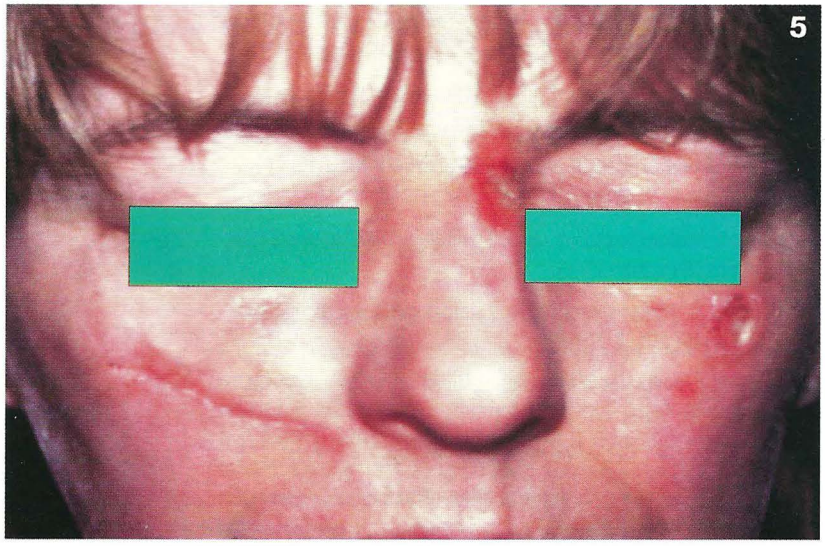
Discussion

Pathomimicry occurs in al fields of medicine. It may aim to obtain a precise material benefit or it may be an expression of psychopathological behavior and, in this second case, it is quite common in intelligent young woman. Since it is important for the practitioner to avoid accusing these patient or attempting to get them to admit to pathomimicry as they are likely to have exaggerated or self-destructive responses, great care must be taken to avoid misdiagnoses. Precisely with this aim, a biopsy was performed in a "difficult" area such as the face of our patient.

In the present case, morphoea exhibited a very unusual and misleading clinical feature: the presence







of ulcerative dermatitis with necrosis. This feature has rarely been reported in Literature and always in the extremities, usually at the extreme tip of the finger, close to the nail (1), or on the lower extremities where lymphatic obstruction combines with increased hydrostatic pressure (3). Out of 53 patients with linear scleroderma, cutaneous ulceration of the involved extremity occurred in only 1 case (4).

Histologically, a precise classification of our case was also difficult. In fact, according to a recent classification (2), the case showed some relationship with morphoea profunda (according to Su and Person (5) as reported by Su and Green in 1986 (6), although in our case inflammation was poor. However, considering the history of blistering reported by the patient and her clinical features as shown in Fig. 5, the case was also related to the bullous subtype, a very rare form since, out of a striking sample of 1071 patients with morphoea and scleroderma only 13 have shown bullous changes (3). Despite their rarity, it is noteworthy that bullae were first described in a patient with morphoea in 1896 (7). The bullous manifestation are considered a consequence of lymphatic obstruction caused by the sclerodermatous process (8-9) or of vascular changes such as arteriitis and phlebosclerosis (10). As further causes of bullous manifestations a localized trauma (6, 11-13) or of the eosinophils, which are often present in cutaneous tissue affected by morphoea (3), can be mentioned.

Considering that the classification of localized morphoea seems to represent a collection of cases with different, although often overlapping, features rather than a precise system based on anatomo-clinical evidence, we have preferred to suggest a new definition for our case, i.e. "deep necrotising morphoea", to highlight its main histological features. We could willfully assign the case one of the mentioned subtypes, though none of these matches in features, but in this case we would not be contributing to advancing the knowledge on morphoea.

Figure 3. This deep arteriole is encircled by collagenous fibers and its wall is hyperplastic (A); the nerve showed in B is also surrounded by the fibrotic tissue (H&E; original magnification X 25).

Figure 4. Necrotic tissue intermingled with black, finely granular foreign material, poorly circumscribed (H&E; original magnification X 10).

Figure 5. Scars on t he face: that in the zygomatico-orbital region is oval and that in the naso-orbital region is rather linear.

The pathogenesis of the ulcerative dermatitis with necrosis in the reported case was amply explained by the striking collagenous deposition not only in the dermis but also in the subcutaneous tissue, which caused lymphatic obstruction and, at a deeper level, arteriolar stenosis complicated by ischemic necrosis of the tissues above. Histologically, bullae were not evident given progression and severity of the disease, but blistering was observed by the patient at onset and the oval ulceration which developed one month after the biopsy looked like an eroded bulla. Pathogenesis of the bullous lesions can be explained by all the mechanisms previously reported in literature. Finally, the involvement of nerves by collagenous deposition observed in our case was in agreement with the peripheral neuropathy previously reported in morphoea (14) and seemed able to exercise a negative neurotrophic effect.

The presence of foreign material exclusively inside the necrotic tissue, without evidence of cellular reaction, confirmed the hypothesis of external contamination. Furthermore, although not convincingly related to the disease in our case, it seems noteworthy that professional exposure to solvents has been previously considered a possible cause of morphoea (15).

Conclusion

The present classification of morphoea appears to be inadequate since it includes a number of insufficiently defined subtypes, which fail to account for all the anatomo-clinical variables, encountered. Such ineffectiveness can lead to serious diagnostic pitfalls, as it happened in our case where exclusion of dermatitis artefacta required a facial biopsy.

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Editorial Note

Deep necrotising morphoea mimicking dermatitis artefacta or dermatitis artefacta mimicking necrotising morphoea?

This editorial note is written in the spirit of open-mindedness and fairness both to the authors and to the readers, because obviously the editors of a journal have a responsibility concerning the scientific and medical content of an article. We disagree with the diagnostic interpretation of the authors based on the study of the clinical photographs and the histopathologic specimens, which were kindly provided by the authors. So, in our interpretation this case represents a dermatitis artefacta somewhat mimicking a morphoea.

Nevertheless, we did not want to reject this manuscript, because the authors were convinced on their interpretation and we like to give them the opportunity to present their concept of deep necrotising morphoea.

The readers are encouraged to decide whether they accept the authors' concept of deep necrotising morphoea or prefer the diagnosis dermatitis artefacta as we do.

Editors

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A U T H O R S ' A D D R E S S E S

Mauro Melato, MD, professor, Institute of Pathological Anatomy of the University of Trieste, c/o Ospedale Maggiore, via Stuparich 1, I-34125 Trieste, Italy
Nader Gorji, MD, same address
Ovidio Marangoni, MD, via S. Francesco d'Assisi 6, I-34100 Trieste, Italy
Giuseppe Pillitteri, MD, via Boccaccio 10, I-34100 Trieste, Italy