Case report

Rosacea with extensive extrafacial lesions

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Abstract

Rosacea is a very common skin disorder in the clinical practice that primarily affects the convex areas of the face. Extrafacial rosacea lesions have occasionally been described, but extensive involvement is exceptional. In the absence of its typical clinical or histological features, the diagnosis of extrafacial rosacea may be problematic. We describe an unusual case of rosacea with very exuberant extrafacial lesions, when compared with the limited involvement of the face.

Introduction

Rosacea is a skin disorder frequently observed in the clinical practice. It is characterized by the primary involvement of convex areas of the face.[1] However, a wide spectrum of clinical findings is often observed.[2] We describe an unusual case of rosacea with exuberant extrafacial involvement.

Case Report

A 53-year-old man presented with an extensive exudative exanthema consisting of papules and erythematous nodules, pustules, and large granulomatous, nodulo-cystic lesions involving the face, neck, and upper chest, and extending from the left shoulder along the whole left arm (Fig. 1). The dermatosis relatively spared the central region of the face, being more exuberant in its lateral aspects and ears. On the upper half of the chest and the left arm (external aspect) the lesions were densely confluent, very inflammatory and exudative, with a crusty surface (Fig. 2). The lesions were pruritic only after sun exposure. The dermatosis had about 6 years of evolution, always localized to the face. The patient reported exacerbation of the lesions, which extended to the chest and left arm, in the past 2 months, following a period of mourning for the death of a member of his family. At this time, he was medicated with oral prednisolone, with transitory improvement of the lesions followed by severe exacerbation. Subsequently, admitting the clinical diagnosis of pustular dermatophytosis, the patient was medicated with itraconazol, with continued worsening of the cutaneous lesions. He had no other pathological antecedents of interest and the remaining physical examination was normal.

Bacteriological and mycological tests of the contents of the pustules were negative. Baseline investigations, including complete blood count, liver and renal functions, autoimmune screen, serology for human immunodeficiency virus, and urine bromides and iodides levels were negative or normal. Phototesting with ultraviolet A (100 J/cm^2 daily) and ultraviolet B (1,5 MED daily) failed to induce the clinical lesions. Histological examination of a lesional biopsies fromy, the face and upper arm, revealed the same features: slight irregular acanthosis, polymorphous inflammatory infiltrate, and formation of epithelioid granulomas in the superficial dermis, and concomitant inflammation of the acute type with formation of follicular pustules (Fig. 3). These findings were consistent with the clinical diagnosis of rosacea. The presence of *Demodex folliculorum* was not noticed.

The patient was treated with deflazacort 30 mg (3 weeks), azithromycin (500 mg, 3 days a week, for 4 weeks), and isotretinoin (10 mg daily, for 1 year, with gradual reduction of the dose in the last months). He was instructed to avoid sun exposure, and a titanium dioxide- and zinc oxide-based sunscreen was prescribed. Gradual resolution of the lesions occurred. One year later, there was still some facial erythema, but complete resolution was achieved on the chest, left shoulder and arm (Fig. 4).

Comment

Usually rosacea is a straightforward diagnosis. However, in the absence of its typical clinical or histological features, the diagnosis of extrafacial rosacea may be problematic. Initially, the clinical differential diagnosis suggested were the many subtypes of infectious folliculitis such as dermatophytosis,

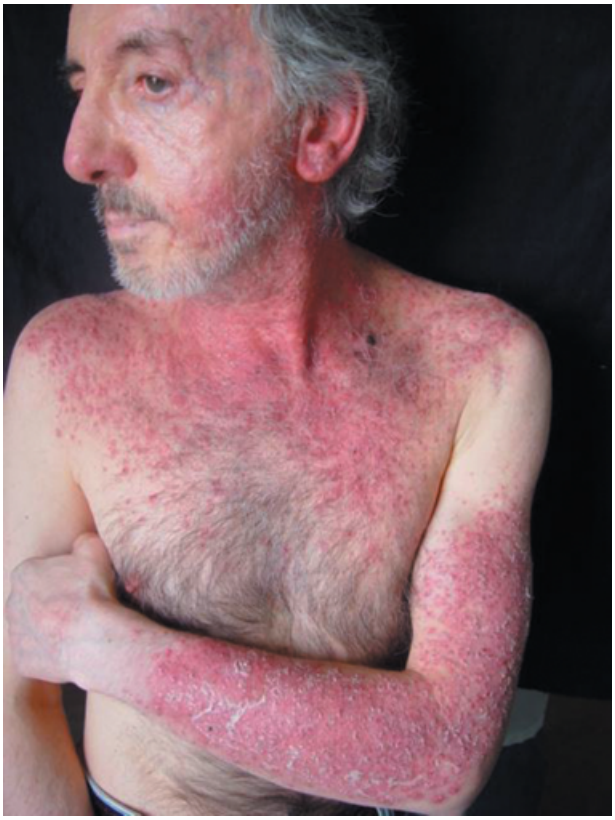

Figure 1 Clinical features of the lesions, with extensive extrafacial involvement

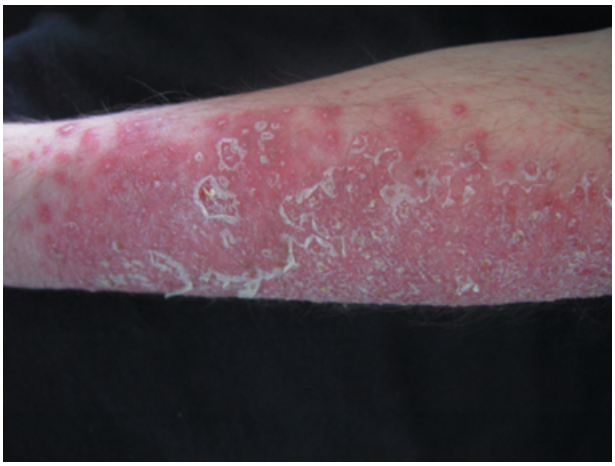

Figure 2 Detail of the lesions on the left forearm

bacterial folliculitis, demodicosis or inflammatory folliculitis, namely, actinic superficial folliculitis, eosinophilic pustular folliculitis, iododerma, or bromoderma. The diagnosis of infectious folliculitis and eosinophilic pustular folliculitis was excluded by the clinical history, bacteriological and mycological tests, and biopsy. Rare patients with actinic superficial folliculitis develop sterile pustules 24–36 h following intense sunlight exposure, with good response to isotretinoin.[3,4] On the contrary, the dermatosis described in our patient showed gradual progression with asymmetric distribution, was not triggered or maintained by sunlight or heat (just caused pruritus with sun) and, inclusively, the phototesting was negative. The patients with halogenoderma (iododerma or bromoderma) tend to develop indurated pustulonodular lesions on the face and extremities, after longer exposure to higher amount of iodides or bromides. Our patient denied ingestion of medications and a measure urine bromides and iodides level was negative.

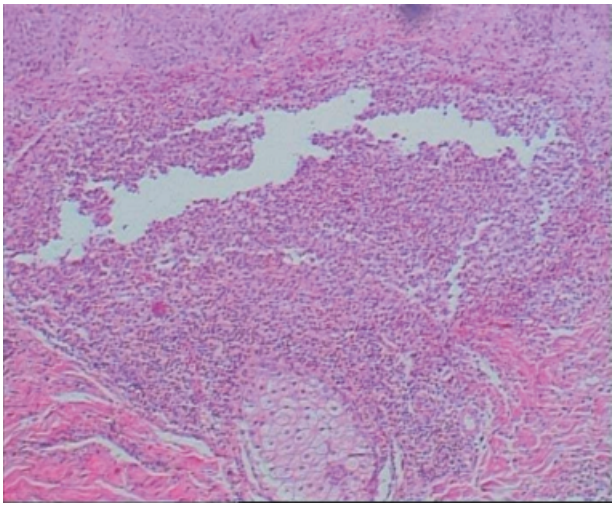

Figure 3 Formation of follicular pustules (hematoxylin and eosin, ×100)

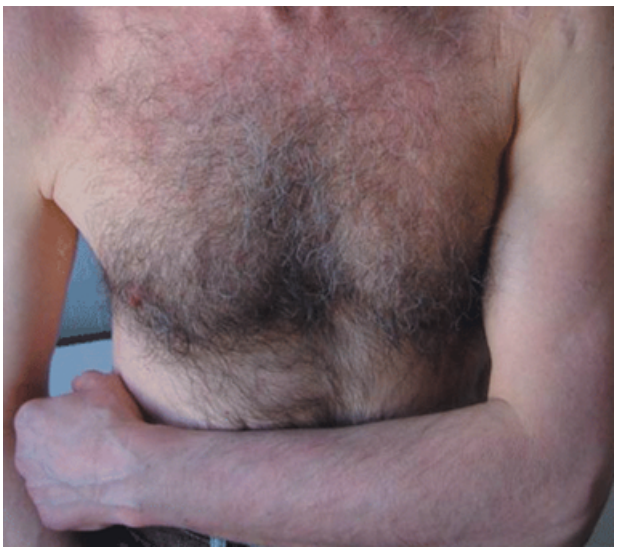

Figure 4 Complete resolution of the lesions after one year of treatment with isotretinoin

Therefore, in this case, the presence clinically of inflammatory papules, pustules, and nodules on a congestive background, with spread from the typical midfacial involvement to other areas of the body, and the presence of poorly organized epithelial granulomas and follicular pustules in the biopsy allow the diagnosis of extrafacial rosacea.

Rosacea is a common skin disorder that is generally localized to the face, more frequently affecting the nose, cheeks, chin, forehead, and glabela, typically sparing the periocular and perioral regions.[2] The presence of rosacea with extrafacial location has been occasionally reported.[5–7] Extrafacial lesions usually complicate the more serious cases of rosacea but, occasionally, as was the case of our patient, they may accompany slight facial lesions. Involvement of the lateral contours of the face, ears, scalp (particularly in old patients), and neck occurs mainly in erythematotelangiectatic rosacea.[2] These extrafacial manifestations are relatively uncommon and generally arise in areas of chronic solar lesions or flushing.[8] Involvement, by acneiform-like lesions, of the chest, axillae, shoulders, arms, and legs (thighs and knees) is described in the literature, but is quite exceptional and generally occurs in the context of the granulomatous variant of rosacea,[5,9,10] as observed in our patient.

Granulomatous rosacea is a variant characterized by papules and monomorphic nodules with erythematous or brownish tonality, generally with periorificial location.[1] Usually, the patient does not have persistent facial erythema.[10] In an anatomoclinical review of 53 cases of granulomatous rosacea, extrafacial lesions were found in 15% of the patients.[11] The authors noticed the presence of those lesions mainly in areas rich in apocrine and pilosebaceous glands, which led them to the conclusion that there is a relationship between granulomatous rosacea and other acneiform eruptions, such as supurative hidrosadenitis.[11]

The cause for the formation of granulomas in rosacea is still unclear. They arise as local macrophage proliferation in response to a persistent antigen, hence, the suggestion by many authors as to the pathogenic role of *Demodex folliculorum*.[12] Ayres,[9] for instance, considers extrafacial rosacea to be caused by a disseminated infestation by *D. folliculorum*, normally inhabiting follicles and sebaceous glands. However, follicular inflammation may be observed in the absence of parasites, and parasites may be observed without associated inflammation.[5,10] A limitative factor in these studies has to do with the fact that *D. folliculorum* is not easily detected in histological preparations: the skin biopsy only shows the parasites that reside superficially in the follicle, but not those located deeper in the follicle or the parasite of *Demodex brevis* residing in sebaceous glands.[2]

Concerning the treatment, we opted for the use of very low doses of isotretinoin (10 mg/day), regardless of body weight, for a long period, with the gradual reduction of the dose to 3–5 days a week. Some studies have established the safety and efficacy of this type of therapeutic approach in difficult cases of rosacea.[13,14] In the long term, cumulative doses above 1075 mg/kg have not caused significant radiological changes.[15] In our patient, the annual cumulative dose is, on average, much lower than that (i.e. 60 mg/kg). The benefit of concomitant short-term corticotherapy has been reported, particularly for granulomatous rosacea.[10] Thus, we also combined isotretinoin with oral corticoids in the first 3 weeks of treatment with good results. In the first month of treatment we also used, azithromycin 3 days a week, in accordance with recent studies that demonstrated the efficacy and safety of this antibiotic in the management of rosacea.[16,17]

Reports of extrafacial rosacea are rather scarce. However, it may be more common than it is thought, because it is generally not considered and/or not researched. We presented an unusual setting of extrafacial rosacea that is remarkable for the exuberant and extensive involvement, when comparing with facial involvement, the initial diagnostic difficulty and the good therapeutic results obtained. This case leaves a number of questions for further study, namely, the potential of extrafacial symptoms in rosacea, the sensitivity to sunlight, the psychogenic factors, and the hypothetical role of *D. folliculorum*.

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