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Facial Granulomatous Dermatoses

Dermatitis granulomatosa facial

To the Editor:

The term *facial granulomatous dermatosis* refers to a group of disorders characterized by facial papules with the common histological feature of epithelioid granulomas. We present a patient whose condition fitted this classification both clinically and histologically, and who illustrates well the difficulties experienced when managing this disorder.

The patient was a 64-year-old woman with no family or personal past history of interest. She was seen in 2005 for asymptomatic facial lesions. On physical examination she presented erythematous papules of 1 mm in diameter in the perioral region, with no comedones or pustules (Figure 1). The initial suspicion was of perioral dermatitis and treatment was started with oral doxycycline (100 mg/12 h) for 3 months, with no improvement. A biopsy



Figure 1 Patient with perioral erythematous papules and pustules. There are no comedones.

was therefore performed; in areas close to the hair follicles there were multiple granulomatous structures formed of epithelioid cells with a peripheral lymphocytic infiltrate and central necrosis, compatible with lupus miliaris disseminatus faciei (LMDf). Ziehl-Neelsen stain performed on the histological specimen was negative for acid and alkali-fast bacilli (Figure 2).

Further additional tests, including complete blood count, erythrocyte sedimentation rate, biochemistry, calcium, Mantoux, chest radiograph, and angiotensin-converting enzyme levels, were normal or negative.

The patient was then administered oral prednisone, with a response only at doses higher than 20 mg/d. She did not tolerate minocycline, sulfone, or thalidomide, and, after oral isotretinoin, developed an intense outbreak of lesions that extended to the periocular and malar regions (Figure 3).

Over time, the outbreaks have affected the periocular region, cheeks, and neck, and, in the last year, have been associated with episodes of facial flushing. Some lesions have left pinpoint scars.

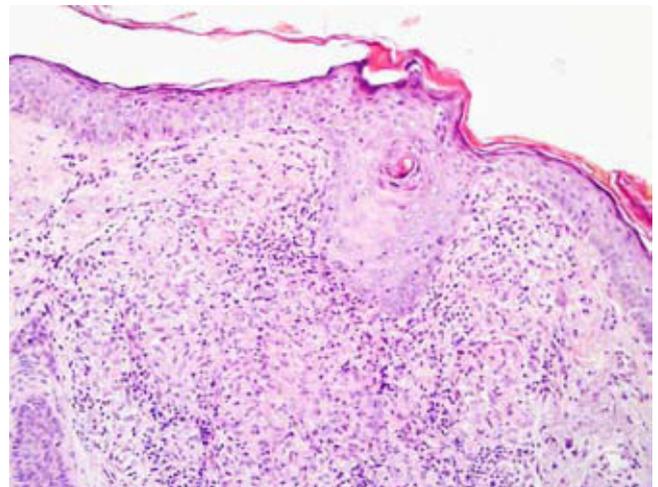


Figure 2 Perifollicular granulomas with epithelioid cells, peripheral lymphocytes, and central necrosis. Hematoxylin-eosin, original magnification $\times 20$.



Figure 3 Frank deterioration with spread to the eyelids and cheeks.

Three years after the initial diagnosis, a further biopsy was performed, showing new epithelioid granulomas related to the hair follicles, some with central necrosis. The pathology report gave a diagnosis of granulomatous rosacea.

Due to the continued therapeutic failure and the intensity of the lesions on the face and around the eyes, it was finally decided to start treatment with oral azithromycin (500 mg/d) 3 times a week. This achieved an immediate response within 2 weeks and that was maintained 7 months later.

LMDF was first described in 1878 by Tilbury Fox, who used the term *disseminated follicular lupus*; since that time it has received many names, including acneitis, acne agminata, and facial idiopathic granulomas with regressive evolution.¹⁻³

It is seen particularly in young adults and is characterized clinically by monomorphic papules situated mainly in the central facial region, with a tendency to affect the eyelids and upper lip, although it can extend onto the neck and nonfacial areas. The papules are yellowish brown and tend to leave pinpoint scars. The condition usually responds poorly to the conventional treatments for rosacea, and often resolves spontaneously in 2 to 4 years.¹

Histology typically reveals the presence of epithelioid granulomas with caseation in contact with follicular structures, but a spectrum of alterations has been described, varying according to the evolutionary phase of the lesion from perifollicular lymphohistiocytic infiltrates to epithelioid granulomas with or without caseation and perifollicular fibrosis in scarred lesions.^{4,5}

Until 1966, LMDF was considered to be a form of cutaneous tuberculosis.⁶ After exclusion of this etiology, the current hypothesis is that it is a granulomatous reaction to follicular components.⁷

Recently, in 2005, a proposal was made to include LMDF in a diagnostic category named facial granulomatous dermatosis, which would also include granulomatous rosacea, perioral granulomatous dermatitis, and the nodular cutaneous form of sarcoidosis.⁸

Although patients with classic rosacea can present a spectrum of histological alterations that include epithelioid granulomas, granulomatous rosacea has been distinguished as a variant of rosacea, and refers to a condition with preferentially periorificial monomorphic papules that can leave a scar and appear on normal skin without other signs of rosacea.⁹ Although granulomatous rosacea responds to the usual treatment for classic rosacea, this definition would situate it as an intermediate disorder between classic rosacea and LMDF.

The clinical presentation of childhood facial granulomatous dermatitis is also different, with perioral lesions in which histology may reveal epithelioid granulomas identical to those described in LMDF. It typically occurs in prepubertal children, particularly black children, and spontaneous resolution is rare.^{10,11}

None of the diseases mentioned above is associated with systemic alterations (fever, general malaise, lymphadenopathies); this aspect differentiates them from cutaneous sarcoidosis, which is considered to be the most extreme of the facial granulomatous dermatoses. Sarcoidosis is, by definition, a multisystem disease, although it has to be said that it often starts with only skin alterations, making diagnosis difficult. It is necessary to exclude other granulomatous diseases, as the diagnosis of sarcoidosis is one of exclusion; there are no definitive diagnostic tests.¹²

Our patient presented the typical features of LMDF, a diagnosis supported by the poor response to tetracyclines and the good response to steroids, in addition to the signs of rosacea (facial redness) that she presented during the course of the disease. All this leads us to consider LMDF, granulomatous rosacea, perioral granulomatous dermatitis, and the nodular cutaneous form of sarcoidosis to be in the same diagnostic category, the facial granulomatous dermatoses.

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Linear Basal Cell Carcinoma

Carcinoma basocelular lineal

To the Editor:

Linear basal cell carcinoma was first described by Lewis¹ in 1985 as a rare morphological variant of basal cell carcinoma. Since that time, 35 cases have been reported in the literature. The most common site is the periocular region.^{2,3}

We report a new case of linear basal cell carcinoma, in which the clinical diagnosis was based on physical examination and dermoscopy and was confirmed by histological study. We review the current literature on linear basal cell carcinoma and discuss its principal features.

The patient, a 42-year-old woman, came to our department for a slowly enlarging, linear pigmented lesion that had appeared one year earlier on the right lower eyelid. She had skin phototype IV. There was no history of trauma or of a previous lesion at that site.

On physical examination there was a linear pigmented lesion measuring 1.5 × 0.4 cm. The lesions had a pearly border and central scarring (Figure 1).



Figure 1 Linear pigmented lesion on the right lower eyelid.

Dermoscopy revealed the absence of a pigmented network pattern and the presence of a specific BCC dermoscopy pattern: multiple blue-gray ovoid nests, arborizing telangiectatic vessels with a feathery appearance, and slight ulceration (Figure 2). The diagnosis of BCC was confirmed by histological study of a punch biopsy (Figure 3A).

The lesion was removed by simple excision, with primary closure of the wound. The surgical margins were free of tumor cells (Figure 3B). Pigmentation was present in the tumor and in the stroma (Figure 3C). After 4 years of follow-up there has been no recurrence.



Figure 2 Dermoscopic aspect.