Morphological Evidence of Periodical Exacerbation of Hyperkeratosis Lenticularis Perstans

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SUMMARY We present a case of hyperkeratosis lenticularis perstans (Flegel’s disease) in a 71-year-old-woman. Apart from all the typical morphological features of the disease, we evidenced mounds of parakeratosis that contained neutrophils, alternating with other strata of the horny layers in which no infiltrate was seen. Such a pattern has been described in association with diseases that show a periodic clinical pattern of presentation with periods of exacerbations and remissions. Our case also showed a lichenoid inflammatory chronic infiltrate with vacuolation of the basal layer and mild spongiosis. Both findings strongly support the hypothesis that the inflammatory infiltrate might play a primary role, and that the hyperkeratosis might be a secondary event.

KEY WORDS: Flegel disease; hyperkeratosis lenticularis perstans; kyrle’s disease

INTRODUCTION

Flegel’s disease (FD) (aka hyperkeratosis lenticularis perstans) is a cutaneous disorder of unknown etiology and pathogenesis (1). Hyperkeratosis is a diagnostic feature of this condition, while dermal inflammatory infiltrate is not a constant and its presence varies in different reports on the disease (2). Some claim that this infiltrate is secondary (2), whereas others claim the opposite (3,4). These discrepancies can be explained if we admit that the disease has a fluctuating course. Some clinical evidence of this fluctuating behavior of FD has already been presented (4,6). Nevertheless, morphological findings supporting such a periodicity have been little emphasized, and when reported, they have mainly been based on the fact that inflammatory infiltrate is absent in old lesions of FD (7). We, therefore, considered interesting to communicate a morphological clue that supports a pattern of exacerbations and remissions of the disease.

CASE REPORT

Our patient, a 71-year-old woman, presented for consultancy, complaining of lesions that had appeared on the posterior side of the forearms, on the shoulders, knees and dorsal aspect of the feet.
for 2 years before. During this period the lesions were asymptomatic. When questioned, the patient reported a protracted course of the lesions. She was taking enalapril due to hypertension and Noctamil to help her sleep. She was allergic to acetylsalicylic acid.

Vacuolization of the basal layer was evidenced (Fig. 4), although no cytoid bodies were seen. The superjacent epidermis showed mild spongiosis. The epidermis was mildly atrophic, with intense hyperkeratosis, mainly orthokeratosis (Fig. 3).

The lesions were oval, with an erythematous halo in many of them (Fig. 1). The biggest lesion measured 0.5 cm. Biopsy was obtained from one of the old lesions of the leg. Morphological study demonstrated a chronic inflammatory infiltrate distributed in a lichenoid pattern as well as in a superficial perivascular pattern (Figs. 2 and 3).

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In the horny hyperkeratotic layer, signs of periodicity exacerbation were easily seen (Fig. 5): they were mainly mounds of parakeratosis that contained neutrophils. They were arranged in different strata, i.e. in a periodic fashion.

DISCUSSION

Flegel's disease is a condition of unknown etiology and pathogenesis (1). The ultrastructural finding of either absence or decrease of membrane coated granules has been claimed as the primary cause and mechanism of the disease (8,9). However, such findings have not been supported by all studies (10,11).
Another morphological feature of the disease is dermal inflammatory infiltrate. The latter damages the basal layer of the epidermis, with evidence of vacuolar change and some apoptotic bodies (3,4,12). Nevertheless, this latter finding is not constant (2), in a way favoring the hypothesis of damage periodicity. This is additionally supported by some recent studies demonstrating differences among FD lesions depending on the time when the biopsy has been taken (7).

In the present case, an important morphological clue was evidenced, i.e. mounds of parakeratosis with neutrophils alternating with either subjacent or superjacent strata of horny layer. This feature has traditionally been associated with diseases in which periods of exacerbation alternate with periods of remission (13).

Although many reports on FD mention focal distribution of parakeratosis in the lesions (3,11), other studies found no parakeratotic mound. For instance, Gutiérrez et al. report on a case of FD only showing compact hyperkeratosis with no parakeratosis (2). Interestingly, their case showed no dermal inflammatory infiltrate either. The absence of inflammation in FD has also been reported in other cases (14), which some consider as a proof that inflammatory infiltrate is a secondary phenomenon (2). Nevertheless, there is no consensus on the latter claim (3,4).

On the contrary, the majority of cases reported in the literature show dermal inflammatory infiltrate (5,6,9,15-21), and it has been demonstrated that old lesions of the disease are mainly those that have no inflammatory infiltrate (7). Some of these discrepancies could be conciliated if the hypothesis of periodic fluctuation of the inflammatory infiltrate is admitted, with periods of exacerbation and periods of remission. The morphological details of the present case would favor such a hypothesis. Moreover, the vacuolar change in the basal layer as well as the lymphocytic epidermotropism could perhaps be the initial signs of aggression to the epidermis, later followed by epidermal hyperkeratotic change.

Nevertheless, an important limitation of our report is the fact that it was a single case.

It is important to remember how clinical fluctuations of Flegel’s disease have previously been reported (4-6). Sometimes a relation between the periods of exacerbation and warm weather (sumertime) has been suggested (4) and ultraviolet light has been considered as a potential provoking factor (5). However, some other cases presented paradoxical spontaneous remission in summer (6).

In conclusion, we present morphological evidence supporting periodical exacerbation of FD, with obvious limitations of a single-case study.

References


Orizol cream - for removing sunspots; year 1929. (From the collection of Mr. Zlatko Puntijar)