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MUCINOSIS FOLLICULARIS PROVOKED BY LIGHT EXPOSURE

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Abstract. The present paper reports a study on a case of follicular mucinosis exacerbated by sunlight exposure. Provocation with standardized light testing was carried out on both normally pigmented skin and on areas of hypopigmentation representing a residual state after earlier skin lesions but without any signs of active mucinosis follicularis. Typical clinical and histologically verified lesions were provoked by the light test procedure, best seen in hypopigmented areas but with similar changes in normally pigmented skin. Characteristic findings developed gradually, assuming an appearance identical with that of the observed spontaneously elicited lesions after 3–4 weeks. Treatment with carotenoids periodically over several years has provided a proper light protective effect, as only minimal lesions have been noted during these periods, compared with extensive changes during similar periods without treatment.

Key words: Mucinosis follicularis; Ultraviolet light, provocation; Light microscopy

Mucinosis follicularis seems to be clearly associated with mycosis fungoides and also other lymphoma, especially in the age group over 40.

The disorder seems to be idiopathic particularly in younger individuals. However, no one seems to have been able to clarify the problem of the etiology or pathogenesis of the disease, although a viral hypothesis had been suggested (7). Johnson et al. (4) proposed some kind of metabolic alteration as an etiological factor. Varying clinical appearances have been described (cf. 2). Usually the lesions are characterized clinically by grouped follicular papules or infiltrated plaques, with loss of hair from the involved follicles.

The histopathological changes of mucinosis follicularis seem initially to engage the pilosebaceous unit, with intercellular oedema. The pronounced accumulation of mucinous material is striking. There is also a more or less prominent concomitant inflammatory cellular infiltrate with perifollicular and periglandular accentuation.

The present paper reports a case of mucinosis follicularis with suspected photo-etiology and experimental light exposure provocation of specific lesions.

METHODS

Case report

The patient is a 22-year-old woman. Two years ago erythematous infiltrated plaques with slight follicular hyperkeratosis appeared on her forehead and cheeks, but also on her arms and legs (Figs. 1 and 2). The lesions partly involved her left eyebrow accompanied by hair loss in this region. There were also vast areas of depigmentation, especially on her upper arms after earlier infiltrations, now clinically healed except for the hypopigmentation.

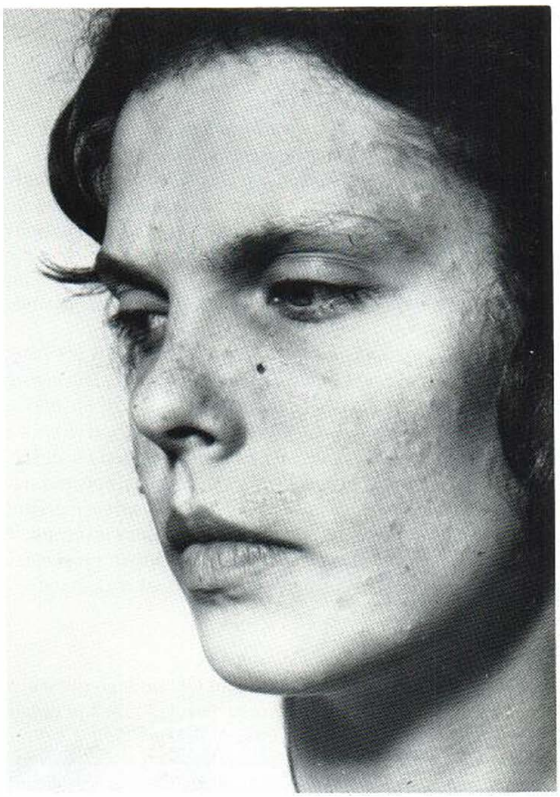

Fig. 1. Skin lesions on forehead and cheek.

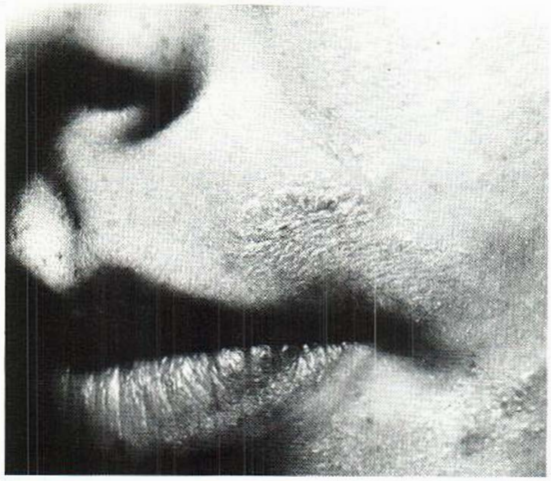

Fig. 2. Detail of the skin lesions.

The lesions were excacerbated by sunlight exposure but there was no general light sensitivity as noted by the patient or found by light testing. She was not taking any medicine, either orally or parenterally and was in good general health and the physical examination revealed no other pathological findings. Routine laboratory examinations, electrophoresis and autoantibody examination, all proved normal.

The patient has been checked at monthly controls for 3 years. Her lesions have regularly faded and disappeared during the winter season, with immediate recurrence following sunlight exposure during alpine vacations.

Treatment with combined carotenoids in a total daily dose of 40 mg beta-carotene and 60 mg canthaxanthine (8) has been given in summertime and on alpine vacations over periods of several months. During these periods the lesions have been noted as slight, compared with her extensive pathological changes during similar time periods without treatment.

Histopathological findings. Histological analyses from a spontaneously developed facial lesion revealed highly characteristic mucinosis follicularis, as described above and as illustrated in Fig. 3. The striking occurrence of mucinous material was histochemically verified by different special staining procedures. The earlier reported accumulation of varying amounts of PAS-positive material could also be demonstrated. The inflammatory infiltrate of the mentioned cellular population was rather preponderant but without signs of cytomorphological malignancy.

Light testing

The light test was performed with an Osram high pressure Xenon arc lamp, XBO 150W, as earlier described in detail (8). The minimum erythema dose (MED) was first established on normal skin of the back for unfiltered radiation. The reactions were read after 8 hr and the dose was noted for the slightest yet perceptible minimum erythema. Afterwards, in order to provoke clinical lesions, a certain skin area was exposed to 8 to 14 MED and this was repeated up to three times on the same area.

The light testing procedure was carried out both on normally pigmented skin and within areas of hypopigmentation representing a residual state after earlier skin lesions but without any clinical or micromorphological signs of mucinosis follicularis.

The light provocation exposure with 8 MED performed three times at intervals of 4–5 days on the hypopigmented area were carried out on two different occasions, but in the second procedure the doses were increased successively, viz. 8, 10 and 12 MED.

Punch biopsies were taken for microscopical analysis at intervals related to the development of macromorphological changes. As a control, a biopsy was perfomed on normal non-irradiated skin.

Parallel with the second light exposure procedure a normal juxtaposed skin area was irradiated with 10, 12 and 14 MED at the same intervals.

The irradiated areas were checked at least once a week.

RESULTS

The MED was established as 5.1 W sec cm^{-2}. Normal MED values in patients without any light-related skin disorders are 2.0 ± 0.6 W sec cm^{-2}. No reactions occurred within the tested skin area when exposed through glass filter up to 132 W sec cm^{-2} as in control patients.

The light provocation exposures elicited the most marked macromorphological changes within the tested depigmented area. After the first light exposure procedure a certain infiltration of the skin appeared after 11 days (Biopsy A 11) which after 27 days (Biopsy A 27) increased and became moderately red and scaly.

A control of exposed hypopigmented skin after 4 days revealed a slight infiltration and an indicated erythema (Biopsy B 4). This reaction gradually intensified (Biopsy B 26) and after 26 days had assumed an appearance identical with the observed spontaneously developed lesions. Within the irradiated normal skin area similar changes appeared after this period (Biopsy C 26).

Histopathology of UV-induced lesions

Biopsy A 11. Slightly hyperplastic epidermis with a moderate oedema, hypogranulosis and corresponding focal parakeratosis and areas of epidermal necrosis. Subepidermally, some perivascular and periappendicular lymphocytic inflammatory infiltration, with deposits of mucin or mucin-like substance as identified with alcian blue and mucicar-

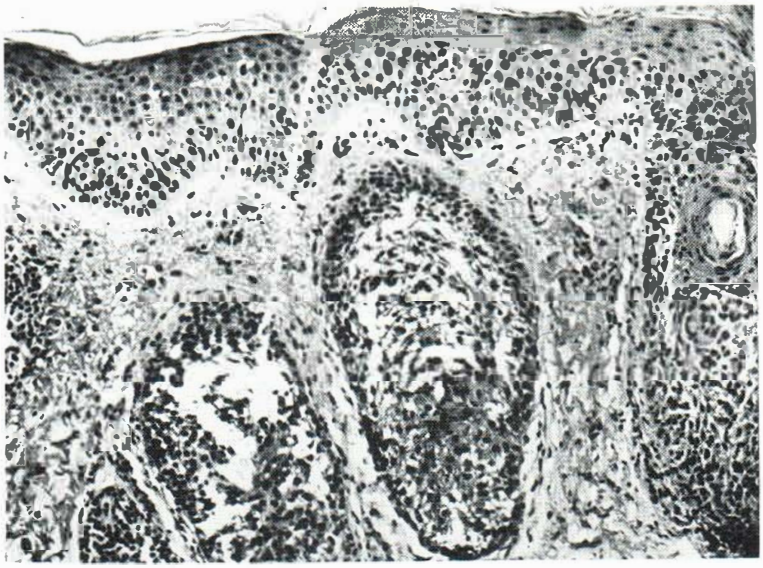

Fig. 3. Occurrence of mucin in the follicles.

mine stains. The findings indicate a developing stage of mucinosis follicularis.

Biopsy A 27 (Fig. 4). Similar histopathological changes but more pronounced, with acanthosis, larger inflammatory infiltrates, except lymphocytes also consisting of numerous eosinophils. Accentuated oedema with marked occurrence of mucin or mucin-like substance within the follicular infundibulum as in mucinosis follicularis.

Biopsy B 26. The histological picture is dominated by an obvious follicular mucinosis.

Biopsy C 26. A moderate perifollicular and intrafollicular oedema with mucinosis material as in the developmental stage of mucinosis follicularis. Sections from the biopsy of normal non-irradiated skin revealed no histopathological changes.

DISCUSSION

The present case of mucinosis follicularis is macromorphologically of the plaque-type, formed through coalescence of follicular papules.

Clinically and partly etiologically, three forms are usually discussed. Many authors recognize a form

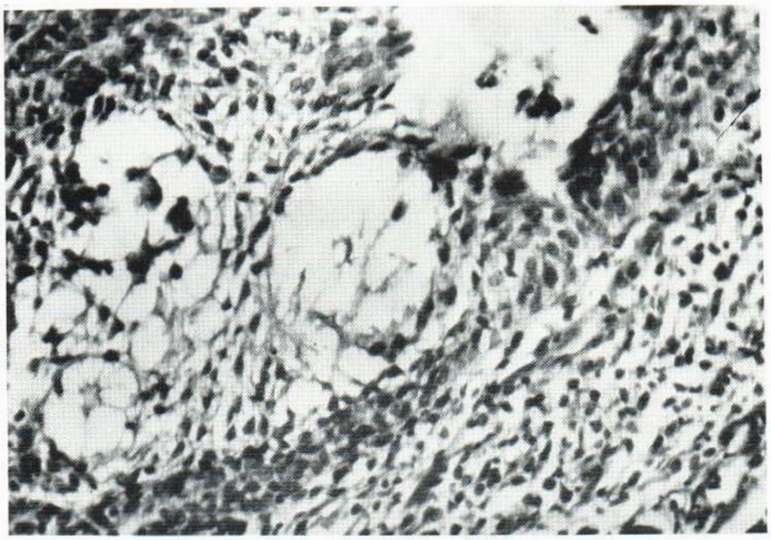

Fig. 4. Part of follicle.

in childhood and adolescence, mostly considered as idiopathic and self-healing after varying periods of manifestation.

Among the adult patients, an idiopathic and a symptomatic form are distinguished. The symptomatic form, especially among patients over the age of forty, is often associated with mycosis fungoides or other malignant lymphomas and may precede this form of malignancy. This association has hitherto appeared to be the only motivation for discussing a symptomatic form of mucinosis follicularis. In the present case, the age of the patient and an observation time of several years seem to contradict any connection with this symptom category.

The present case of mucinosis follicularis is not of the idiopathic type, as was proven by the light testing procedure and by microscopical analyses.

Typical clinical and histologically verified lesions of follicular mucinosis were in this case spontaneously induced by solar irradiation and provoked by the light testing procedure. This marked effect of UV light has not been reported previously.

As a control, 15 patients with similar elevated MED values as in the present case were provoked by a similar light testing procedure. In this control group no characteristic macro- or micromorphological changes of mucinosis follicularis appeared.

This investigation definitely indicates the existence of a special type of symptomatic follicular mucinosis which is induced by UV light.

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