CORRESPONDENCE

Mycosis fungoides with xanthomatization

Case report

A 37-year-old man presented with a 10-year history of brownish patch on his right upper arm with yellowish discoloration developing gradually in the past 2 years (Figure 1A). In addition, another two brownish patches on his left flank and right thigh were first observed 6 and 4 years ago, respectively. Physical examination revealed three discolored patches without definite scaling and ulceration over the right upper arm (6 × 4 cm, brownish to yellowish), the left flank (5 × 4 cm, brownish), and the right thigh (4 × 3 cm, brownish), respectively. Laboratory tests showed an elevated fasting cholesterol of 5.73 mmol/L (3–5), a fasting triglyceride of 2 mmol/L (0.6–1.2), and a low-density lipoprotein of 4.3 mmol/L (2–3), whereas the high-density lipoprotein was normal at 1.1 mmol/L (0.9–2). A follow-up examination was performed 1 year later, and it revealed a fasting cholesterol of 5.43 mmol/L (3–5), a fasting triglyceride of 1.94 mmol/L (0.6–1.2), a low-density lipoprotein of 3.8 mmol/L (2–3), and a high-density lipoprotein of 1.1 mmol/L (0.9–2). The skin biopsy from the brownish patch on the flank revealed the typical dermatopathology of mycosis fungoides (MF) at the patch stage, and the biopsy from the brown-yellowish patch over the upper arm showed epidermotropism, and below the lymphocytic infiltration were many foamy cells in close proximity (Figure 2A–D), which stained positive for CD68 (Figure 3C) but negative for CD45RO. Most epidermotropic atypical lymphocytes were stained positive for CD4 and only a few positive for CD8 (Figure 3A,B). The patient received regular narrow band UVB (NBUVB) phototherapy for 1 year (dose = 0.5–1.6 J/cm², cumulative dose = 172.75 J) and the lesion has diminished significantly (Figure 1B).

Discussion

Cutaneous T cell lymphoma (CTCL) is composed of a heterogeneous group of malignant lymphomas. MF is the most common variant of CTCL, and the typical clinical manifestation of MF is erythematous macules and papules at the early stage, often resembling eczema or psoriasis.

We should differentiate this case clinically from granulomatous slack skin (GSS) because GSS may also involve the area of axilla. GSS is clinically manifested as red to violet, atrophic and well-circumscribed plaques with fine scales over intertriginous areas, such as axilla and inguinal areas. The dermatopathological features of GSS are small- to medium-sized lymphocytes in the dermis, non-caseating granulomas, histiocytes and multinucleated cells containing very large numbers of nuclei throughout the dermis to the subcutaneous tissue. Thus, our case is easily distinguished from GSS by pathological findings.

To date, xanthomatous change in MF has been rarely reported. Xanthomatous change may occur in tumor stage MF as a result of dystrophic change. McCadden et al has proposed that cellular lipids were released from the neoplastic cells after local tissue hypermetabolism or damage following therapy, and then the lipids were processed and engulfed by macrophages. In another report, Ross et al have reported a case of MF with xanthomatized atypical T lymphocytes in a subject with hyperlipidemia. It was suggested that the malignant T cell might play a direct role in the processing of lipoprotein and the xanthomatization. In our case, although there was significant improvement of the lesions after a course of 1-year NBUVB phototherapy, the blood lipid levels were slightly higher than the normal ranges both before and after the course of

Figure 1 (A) The brownish patch with yellowish discoloration over the right upper arm showed no definite scaling and ulceration. (B) A significantly regressed macule after NBUVB treatment for 1 year was noted in the same lesion.
treatment. Thus, we suspected that the pathomechanism of xanthomatization might not be related to the patient’s hyperlipidemia. Moreover, these xanthomatized cells stained positive for CD68 and negative for CD45RO, suggesting that these cells are macrophages rather than T cells. Taken together, this finding suggests that the xanthomatization may be the result of local dystrophic change.

Xanthomatization in both inflammatory and neoplastic cutaneous diseases has been previously reported. In CTCL, xanthomatization has been shown to occur as a result of dystrophic change or to be secondary to systemic hyperlipidemia. However, xanthomatization occurred exclusively at the tumor stage of CTCL in all the previous reports.

The therapy of early stage MF has various modalities, including topical potent steroids, topical nitrogen mustard, topical camu camu, psoralen with ultraviolet A (PUVA), NBUVB, and electron beam radiotherapy. The mechanism of action of NBUVB for MF still remains unclear, but the possible explanation includes inhibition of the antigen-presenting function of Langerhans cells, induction of the apoptosis and reduction of the proliferation of clonal T cells.

Here we report for the first time that xanthomatization was observed in the patch stage MF, but the exact pathomechanism of the foamy histiocytes at the patch stage of MF still remains elusive and awaits further study.

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References