Case Report

Leser-Trélat Sign Presenting in a Patient with Relapsing Mycosis Fungoides

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Keywords
Leser-Trélat sign · Mycosis fungoides · Paraneoplastic · Seborrheic keratoses

Abstract
The Leser-Trélat sign is a rare sign of some malignant tumors and is characterized by the sudden appearance of seborrheic keratosis in association with an underlying malignancy. We describe a 60-year-old Saudi man with mycosis fungoides (MF) who developed numerous, rapidly growing, seborrheic keratoses on his face and back. To the best of our knowledge, this is the first reported case of MF with the Leser-Trélat sign from Saudi Arabia.

Introduction

The sign of Leser-Trélat (LT) is characterized by the abrupt appearance of multiple seborrheic keratoses, sometimes associated with pruritus in association with an underlying malignancy [1, 2]. It has been reported as a paraneoplastic disease in the setting of various types of malignancies, most commonly adenocarcinoma of the stomach [3, 4]. However, a few cases of mycosis fungoides (MF) with LT sign have been reported [5–9]. To the best of our knowledge, no cases of LT sign have been reported among the Saudi population.
Case Report

A 60-year-old male presented to the Dermatology Clinic at the King Khalid Hospital on 2007 with a 3-year history of itchy, confluent, erythematous, mildly scaling macules and patches on his trunk and extremities. The body surface involvement was around 30%. The physical examination was otherwise unremarkable. Histopathological evaluation revealed features consistent with MF. Investigations (complete blood count [CBC], liver function [LFT], urea and electrolyte [U/E], lactate dehydrogenase [LDH], peripheral blood flow cytometry tests and CT of chest, abdomen, and pelvis) were all normal.

A diagnosis of patch-stage MF (stage IB; TII, N0, M0) was made. The patient was started on treatment with narrowband ultraviolet B 3 times weekly for 9 months with complete clearance, and skin biopsy showed no evidence of MF. The follow-up showed a lasting remission of the disease over the next 5 years; then he missed his follow-up for 3 years. In March 2017, the patient presented to our clinic with progressive itchy erythematous patches on the trunk of 9 months’ duration, and he had noted the sudden appearance of multiple black-colored lesions over his face and trunk.

On examination, there were multiple erythematous scaly plaques on his abdomen and thighs, the body surface involvement was around 8%, and there were numerous seborrheic keratoses on his face, upper limbs, and trunk (Fig. 1, 2). There was no lymphadenopathy or organomegaly.

A biopsy specimen taken from an erythematous plaque on his left lower back showed focal parakeratosis and mild epidermal hyperplasia. There was superficial lymphocytic infiltrate in the superficial dermis with epidermotropism of few atypical looking cells. By immunohistochemistry stains, these cells are positive for CD3 and CD4 with a marked decrease in CD7 and CD8. Another biopsy taken from his upper back showed acanthosis, pigmented keratinocytes, and pseudohorn cysts consistent with seborrheic keratosis (Fig. 3, 4).

A comprehensive investigation panel, including CBC with differential, LFT, U/E, and LDH, was normal. Flow cytometry of the peripheral blood was normal, and the CD4/CD8 ratio was normal. CT of chest, abdomen, and pelvis was done and showed no lymphadenopathy or organomegaly.

Discussion

The sign of LT is characterized by a sudden eruption of many new seborrheic keratoses or a rapid increase in their size over a short time (weeks or months), often associated with pruritus and primarily affecting the upper trunk and the dorsum of hands, followed by extremities, face, abdomen, neck, and axilla [10]. As suggested by Heaphy et al. [11] the “syndrome of Leser-Trélat” is defined as a paraneoplastic syndrome in patients with the “sign of Leser-Trélat,” in whom an occult malignancy is identified after the appearance of the sign. However, detecting the sign upon history and physical examination alone with or without the association with occult malignancy can be referred to as the “sign of Leser-Trélat.”

LT is predominantly associated with solid organ carcinomas and rarely occurs in association with hematopoietic malignancies, especially cutaneous T-cell lymphoma; only 10 cases of the LT sign have previously been reported in the setting of MF [8, 9].

In this report, the patient initially presented with itchy, confluent, erythematous, mildly scaling macules and patches on his trunk and extremities, representing the patch-stage MF (stage IB; TII, N0, M0). This was followed by the multiple black-colored lesions over his face.
and trunk 10 years after confirming his diagnosis of MF. This was different from a case reported from Japan [7], where the patient developed brownish papules on the front of her neck and chest only a few weeks before the development of the generalized erythematous eruption. In another case reported by Narala et al. [8], the patient noted several brown skin lesions concurrent with a red, scaly rash over the entire body, which could be related to the fact that the patient did not comply with his appointments, leading to a late diagnosis. Also, the differences in the timing of LT sign presentation in relation to MF lesions indicate that it is not associated with the aggressiveness or stage of MF.

Miyako et al. [7] suggested a specific LT sign pattern that could be linked to an MF-associated LT sign. They noted that LT sign did not develop in a typical way; seborrheic keratoses appeared only on the neck and chest, and they did not further increase in size. In our patient, LT sign had developed over his face and trunk, questioning the suggested pattern of MF-associated LT sign.

Conclusion

Seborrheic keratoses are common findings in elderly patients. Therefore, patients with a sign of LT should undergo a diagnostic screening program for malignant disease. Moreover, these lesions may coexist with the diagnosis of different types of cancers, or follow or precede it by months or years.

Statement of Ethics

Patient informed consent was signed by the patient.

Disclosure Statement

The authors report no financial and personal interests and have no conflicts of interest.

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Fig. 1. Clinical picture of the eruption of the seborrheic keratosis over face.
Fig. 2. Clinical picture of the eruption of the seborrheic keratosis over back and multiple lesions of MF.

Fig. 3. Upper back lesion reveals acanthosis, pigmented keratinocytes, and pseudohorn cysts consistent with seborrheic keratosis. H/E stain. ×100.
Fig. 4. Biopsy from right lower back exhibits atypical lymphocytes in epidermis with papillary dermal fibrosis. H/E stain. ×400.