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Research Article

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Acral Lentiginous melanoma arising in Brauer-Buschke-Fischer plantar keratoderma

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Abstract

Type I Punctuate palmoplantar keratoderma or Brauer-Buschke-Fischer keratoderma (BBFK) is a rare autosomal dominant keratinisation disorder with variable penetrance. \r\n The emergence of acral lentiginous melanoma on palmoplantar keratoderma is rarely reported. Herein, we present a case of a 53-year-old woman with a history of BBFK in both palms and soles, who developed an acral lentiginous melanoma. The clinical examination showed multiple tiny hyperkeratotic and translucent papules on both palms and soles. On the right sole, she developed an acral lentiginous melanoma over a hyperkeratoic area. This case presentation illustrates a rare association of malignant melanoma and BBFK and seeks to enhance further investigations to determine its pathophysiological mechanism.\r\n

Introduction

Brauer-Buschke-Fischer Keratoderma (BBFK) is an autosomal-dominant inherited condition belonging to the group of punctuate palmoplantar keratoderma (PPPK) [1]. It is rarely associated with malignancies. We report a case of acral lentiginous melanoma (ALM) of the right sole arising on a non-transgradient PPPK.

Case presentation

A 53-year-old agriculturist Tunisian woman was admitted for a pigmented lesion of the right sole, evolving for few years. Her past medical history included painful palmoplantar keratoderma since the age of 30 which progressively worsened over the years. The patient was not regularly followed-up for her keratoderma. She recently reported loss of weight and appetite.

Skin examination revealed numerous translucent and tiny orange-yellowish papules of different size (1-4mm) with irregular distribution in both palms and soles (fig1a,b,c).

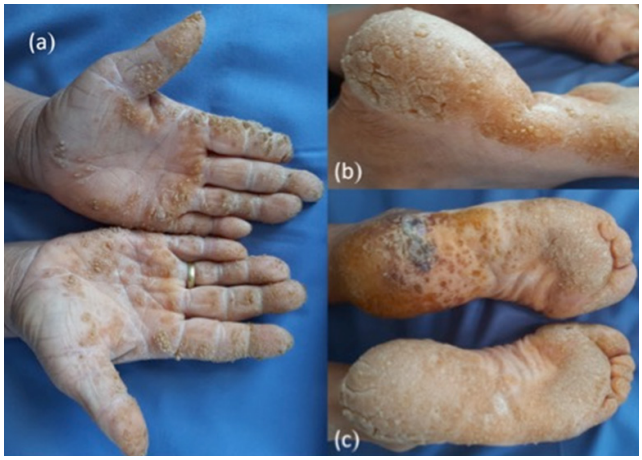

Figure 1a, b, c: Buschke Fisher palmoplantar keratoderma presenting as multiple and confluent keratotic papules over the palms and soles (a,b) with associated pigmented and nodular lesion over the right sole (c).

The lesions were confluent in rough patches mostly located in pressure areas. On the right sole, a 3cm-heterochromatic ulcerated and nodular tumor was noticed over a keratotic plaque. Physical examination revealed no abnormalities except a 7,5x4cm firm and painful mass in the right inguinal region. Dermoscopy of plantar lesions showed an ulcerated pigmented tumor with parallel ridge pattern, polychromia, blue-white veil and asymmetric structureless areas (fig2a,b), which was surrounded by keratotic and translucent papules with central depression (fig2c).

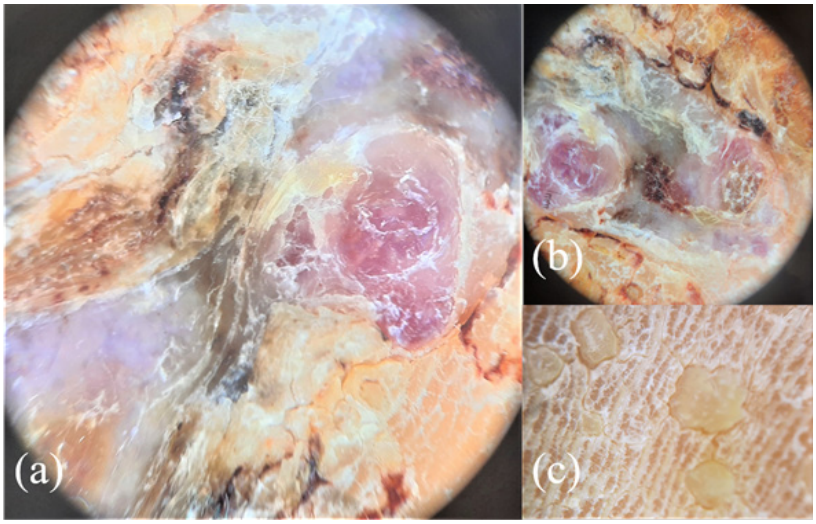

Fig2a, b, c: Dermoscopic examination showing a pigmented plantar tumour with parallel ridge pattern and chaos of colors and structures (a,b) surrounded by keratotic papules with central depression (c).

Histological examination of keratotic papules showed orthokeratotic hyperkeratosis, hypergranulosis, acanthosis and epidermal depression while the analysis of the pigmented lesion (fig. 3a,b) showed confluent epithelioid melanocytes along the dermal-epidermal junction and pagetoid spread of atypical melanocytes with high mitotic rate (40/10). The diagnosis of BBFK with associated ALM was made. Mutation analysis of AAGAB gene could not be performed. The results of laboratory tests including lactate dehydrogenase and tumor markers blood levels were within normal limits. The ultrasound of the inguinal lymph node identified clear aspects of malignancies. Lymphadenectomy was performed and anatomopathological examination was positive for metastasis. A whole-body computerized tomography scan demonstrated no evidence of distant metastasis. The tumor was completely removed with 3cm margins. The Breslow thickness was 5mm and Clark level was IV.

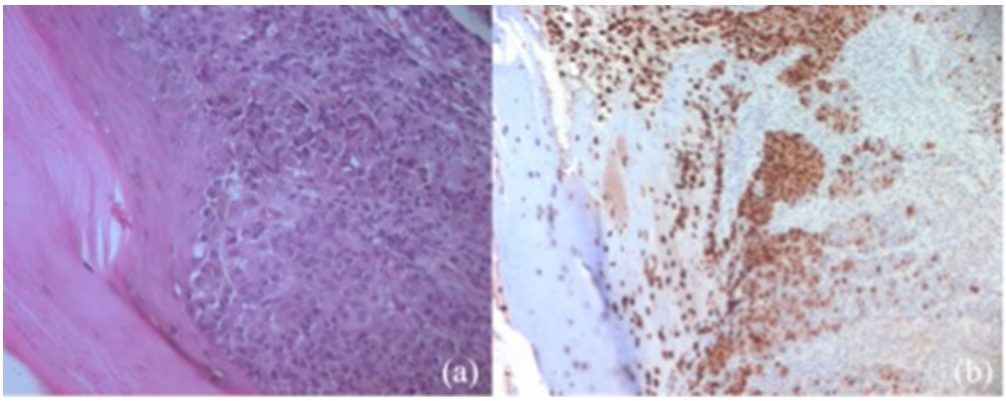

Fig3 a, b: Histological examination of the pigmented lesions showing epithelioid melanocytes along the dermal-epidermal junction and pagetoid spread of atypical melanocytes with hyperchromatic nuclei, atypical mitoses and high mitotic rate (40/10).

The ALM stage was pT4bN1bM0, stage IIIC

The residual defect was managed by directed wound healing with satisfying functional and cosmetic results. The Palmoplantar keratodermas are divided to four groups: diffuse, focal, punctuate and striate. BBFK represents type I in the classification of PPPK due to mutations in AAGAB gene[1]. BBFK occurs as multiple hyperkeratotic papules of different size involving the palms and soles. BBFK is rarely associated with malignancies.

ALM is a rare subtype of malignant melanoma (MM) with infrequent BRAF mutations [2].

Like other acral malignant tumors, ALMs have significant diagnostic delay, related to their hidden anatomic site. This delay can be even worse in patients who have diseases such as keratoderma. A regular clinical and dermoscopic follow-up of the plantar keratoderma in our patient, would have allowed an early diagnosis of ALM and improved its prognosis.

Conclusion

We report a case of ALM arising in plantar BBFK, with delayed diagnosis ans poor prognosis. Regular clinical and dermoscopic follow-up is crucial to detect suspicious acral pigmented lesions, especially in patients with palmoplantar keratoderma.

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