Acral Melanoma in an Elderly Patient with Congenital Ichthyosis Vulgaris

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Received: February 1, 2022
Accepted: September 1, 2022

ABSTRACT Ichthyoses are a heterogeneous group of skin disorders featuring erythroderma and generalized scaling. The relationship between ichthyosis and melanoma has not been well-characterized. Herein we present a unique case of acral melanoma of the palm developing in an elderly patient with congenital ichthyosis vulgaris. Biopsy revealed a superficially spreading melanoma with ulceration. To the best of our knowledge, no acral melanomas have been reported so far in patients with congenital ichthyosis. Nevertheless, considering the potential for invasion and metastasis, patients with ichthyosis vulgaris should undergo regular clinical and dermatoscopic screening for melanoma.

KEY WORDS: acral melanoma, ichthyosis, dermatoscopy, screening, skin cancer

Competing interests:
The authors received no financial support for the research, authorship, and/ or publication of this article. The authors declare no conflict of interest. Only records of patients who had given informed consent for the acquisition of photographic images and medical data were included.

INTRODUCTION

Ichthyoses are a heterogeneous group of skin disorders featuring generalized scaling. The relationship between ichthyosis and melanoma remains largely unknown. Herein, we present a unique case of acral melanoma developing in an elderly patient with congenital ichthyosis vulgaris.

PATIENTS AND METHODS

A 78-year-old woman presented with a nine-month history of a progressively enlarging, friable, pigmented lesion on her left palm that bled easily after minor trauma. The patient had been diagnosed with congenital ichthyosis vulgaris during infancy. Her family history was negative for both ichthyosis vulgaris and melanoma. Clinical examination revealed mild erythroderma and dry, scaly skin with detachable white to grey scales, acral hyperlinearity, ectropion, and bilateral hallux valgus. On the left palm, a large, bleeding, brown-black lesion with irregular borders was noted (Figure 1, A). The dermatoscopic examination showed multiple colors (brown, black, red, grey, blue), a 'rainbow pattern', parallel ridge pattern, ulceration, dotted and linear vessels and blue-white areas (Figure 1, B, C).

A partial excision of the nodular part of the lesion revealed a superficially spreading melanoma with a Breslow thickness of 1.8 mm, low mitotic rate, and ulceration (Figure 2). The patient was instructed to undergo re-excision and sentinel lymph node biopsy. However, she missed surgical and oncologic appointments and was subsequently lost to follow-up.

Acta Dermatovenerol Croat
2022;30(3):185-187

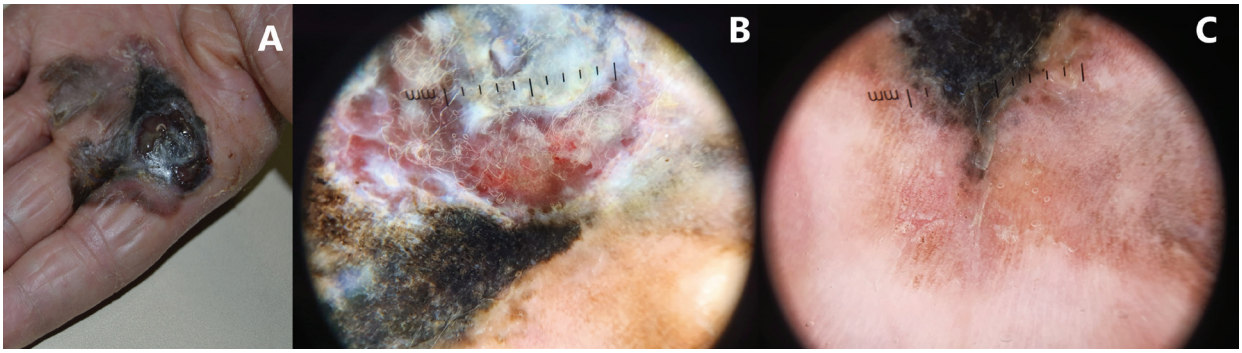

Figure 1. Clinical and dermatoscopic findings. A) A melanocytic lesion on the palm of the left hand with a central friable nodular part. (B) Dermatoscopic image featuring multiple colors (brown, black, red, grey, blue), 'rainbow pattern', a parallel ridge pattern, ulceration with adherent fiber, dotted and linear vessels, and blue-white areas. (C) Dermatoscopic image showing a parallel ridge pattern, asymmetric distribution of color, an asymmetric black blotch, and regression areas.

DISCUSSION

Ichthyoses can be limited to skin involvement or be associated with systemic manifestations. Both inherited and acquired forms have been described (1). Depending on the type of ichthyosis and individual and environmental factors, clinical presentation may range from mild xerosis and scaling of the extremities to generalized, massive hyperkeratosis and scaling (2). Over fifty genes have been linked with inherited ichthyoses (3,4). Ichthyosis vulgaris is the most common disorder of keratinization, estimated to affect 1 in 250 individuals (3-5). It is caused by loss-of-function mutations in the filaggrin gene and is inherited in an autosomal semi-dominant fashion with 83-96% penetrance (3,6,7). Clinical features include grey-white scales on an erythematosus background, palmoplantar keratoderma, nail dystrophy, alopecia, ectropion and anhydrosis (8). Ichthyosis vulgaris is associated with an increased risk of immediate hypersensitivity reactions such as bronchial asthma, drug allergies, and atopic dermatitis (4,5).

Several reports showed a predisposition for skin cancer in patients with congenital ichthyoses (6,9-11). Damage to the epidermal barrier, loss of pigmentation, and incomplete maturation of epidermal cells due to their increased mitotic rate have all been suggested to increase the risk of skin cancers (9,11). The most common skin malignancies associated with various types of congenital ichthyoses were cutaneous squamous cell carcinomas (6,9-11). Incidence of basal

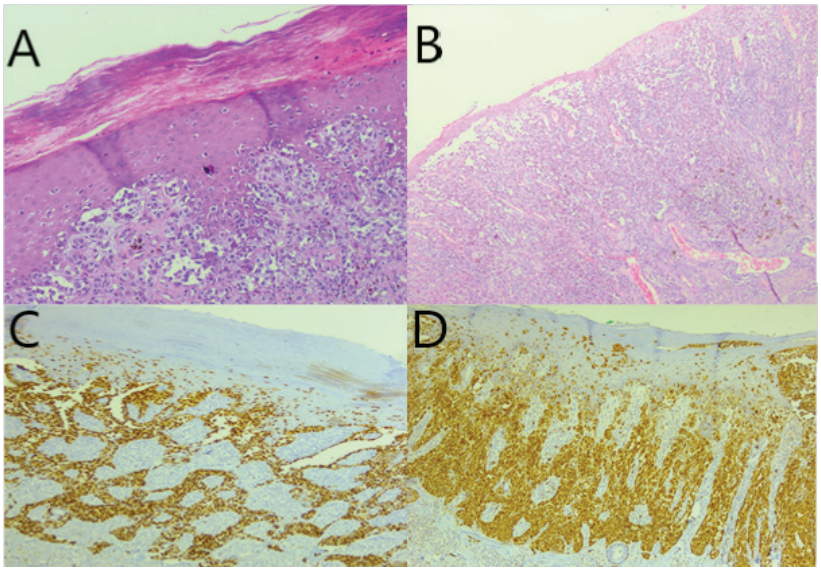

Figure 2. Histopathological images. (A) Hematoxylin-eosin staining, magnification ×5. Superficially spreading melanoma featuring proliferation of atypical melanocytes with pagetoid migration. Tumor cells are medium to large in size, with abundant eosinophilic cytoplasm and large, irregular, atypical nuclei with abundant eosinophilic nucleoli. (B) Hematoxylin-eosin staining, magnification ×5. Superficially spreading melanoma featuring tumor cells with ulceration of the surface epithelium. (C) Tumor cells are positive for Sox-10 marker (magnification ×5). (D) Tumor cells are positive for antigen S100 (magnification ×5).

cell carcinomas and malignant proliferative trichilemmal tumors also appeared to be increased in patients with congenital ichthyoses (12-16). Cases of cutaneous lymphoma and malignant fibrous histiocytoma have also been reported (11,17). So far, cutaneous melanoma was described in four cases, all featuring patients with non-bullous congenital ichthyosiform erythroderma with no apparent risk factors for melanoma (6,9,11). Clinically, they all presented with ulcerated, bleeding nodules, two of which were visibly pigmented lesions (6,11) and the other two were papulo-erythematosus lesions (9,11). In all cases, melanoma involved the extremities: one was located on the shoulder (6), another one on the forearm (9), and two on the thighs (11). Biopsy of the lesions revealed superficially spreading melanomas (6,9).

CONCLUSION

To the best of our knowledge, no acral melanomas have been reported so far in patients with congenital ichthyosis. This can be explained by the rarity of the two afflictions and it makes our case unique. Nevertheless, considering the potential for invasion and metastasis, we believe that patients with ichthyosis vulgaris should undergo regular clinical and dermatoscopic screening for melanoma.

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