

Rapidly fatal metastatic melanoma arising from a congenital nevus in a young female

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Sir,

Melanoma is an aggressive and potentially fatal tumor of melanocytic origin. It may occur at any age yet more rarely at a young age [1]. Melanomas in young patients have overall a more favorable prognosis than in older. However, progression to the metastatic stage and the death of the patient are not exceptional [2]. One of the major risk factors for the development of melanoma in children and young adults is the congenital melanocytic nevus (CMN) [3]. The risk of malignant transformation of all congenital nevi ranges from 0.05% to 10.7%. The risk of malignant degeneration is correlated with size and location [4]. The size of the CMN above 40 cm as well as the presence of satellite nevi and the location in the trunk seem to increase the risk of developing MM. The role of surgical removal in inducing melanomas is controversial. In anatomopathology, melanomas arising from CMNs are usually located in the dermis and hypodermis, while melanocytic proliferation in a melanoma without a CMN starts in the epidermis [3]. Given the differences in the anatomical involvement of the disease, melanoma arising from congenital nevi may be considered a separate entity from the conventional case of melanoma and management may differ. Large excision may not be sufficient to remove all neoplastic cells from the nevi, and adjuvant aggressive systemic therapies may be essential to avoid a fatal outcome [4]. A recent study revealed that congenital nevi preferentially harbor NRAS mutations rather than BRAF mutations commonly seen in other types of nevi, indicating an altered molecular basis of nevocgenesis in congenital nevi [3]. Herein, we report the case of a

rapidly fatal metastatic melanoma in a young female arising from a congenital nevus of the trunk.

A young female 24 years of age presented with a pigmented, congenital, 5 cm lesion of the abdomen. The patient underwent surgical excision of the lesion without histological assessment. A painful angiomatic nodule appeared over the existing lesion evolving for the last year (Fig. 1). The evolution was then marked three months later by the appearance of numerous erythematous and angiomatic cutaneous and subcutaneous nodules disseminated over the entire body (Figs. 2a and 2b). We also noted inguinal and axillary bilateral lymph nodes associated with asthenia, dyspnea, headaches, and dizziness. A biopsy of the angiomatic nodule adjacent to the nevus



Figure 1: The pigmented angiomatic nodule above an asymmetric pigmented macule in the abdominal area.

How to cite this article: Kassel J, Elloudi S, Soussy K, Chhiti S, Baybay H, Douhi Z, Bouhafa T, Mernissi FZ. Rapidly fatal metastatic melanoma arising from a congenital nevus in a young female. *Our Dermatol Online*. 2023;14(1):119-120.

Submission: 06.01.2022; **Acceptance:** 03.03.2022

DOI: 10.7241/ourd.20231.33



Figure 2: (a) Multiple subcutaneous nodules on the back. (b) The angiomatous nodule at the abdominal level.

was performed and showed a predominant dermal proliferation with some intraepidermal nests of atypical melanocytic cells with foci of tumor necrosis and ulceration. IHC showed a positive marking of

HMB45 and Melan A. A total body CT scan was performed showing metastases of the brain, lungs, soft tissue, pancreas, and bone with peritoneal calcinosis. The patient was transferred to the oncology and radiotherapy department and deceased two weeks later.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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Source of Support: Nil, Conflict of Interest: None declared.