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Whitish halo on a papular pigmented lesion

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A 49-year-old healthy Caucasian woman presented with a sudden change in a lesion on her abdomen. At the time of our consultation, we observed a pigmented papular lesion, 1.5 cm in diameter, surrounded by an incomplete whitish halo with two dark rounded blotches at its top (Figure 1a).

The patient had a fair skin phototype and chronic photoaging, characterized by atrophic skin, wrinkling, and diffuse solar elastosis, especially on the sun-exposed areas. Dermoscopy showed a brownish irregularly oval plaque with a few dotted vessels and regular globules. On top of the lesion, two heavily asymmetric and pigmented blotches were visible: on the left, it was bluish and on the right side reddish (Figure 1b).

Histopathological examination revealed a compound, predominantly intradermal melanocytic nevus with congenital features adjacent to a focal scar, suggestive of a previous trauma or surgical procedure (Figure 2a). Within the lesion, an expanding dermal nodule was detected, characterized by nests of epithelioid melanocytes. No nuclear cell atypia, pleomorphism, mitosis, or pagetoid spread into the epidermis was observed (Figure 2b). The nodule was characterized by large melanocytes with minimal pleomorphism, without evidence of necrosis (Figure 2c). The periphery of the nodule showed maturation of the melanocytes blending with the surrounding nevus.

Cells of the congenital nevus and cells of the dermal nodule were positive for S-100 protein and negative for HMB45. The Ki-67 proliferation index was low in the congenital nevus but moderately high in the nodule, while p-16 was negative in the nodule, but positive in the remainder of the nevus. BAP-1 immunohistochemistry revealed BAP-1 nuclear staining (Figure 2d).

What is your diagnosis?

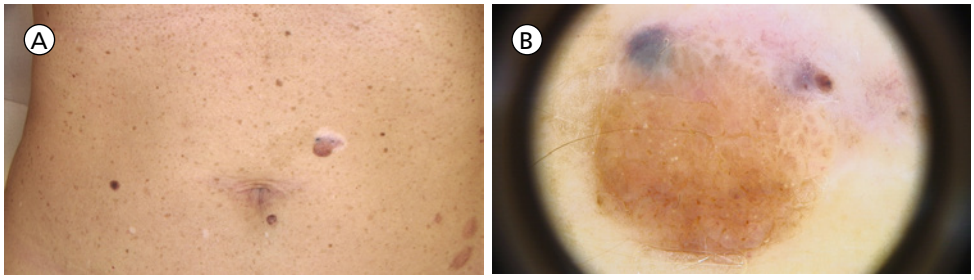

Figure 1. (a) Clinical presentation. Papular pigmented lesion measuring 1.5 cm in diameter, surrounded by an incomplete whitish halo and areas of hyperpigmentation on its top. (b) Dermoscopic images revealed a regular pigmented network with two hyperpigmented blotches on its top, one bluish and one reddish, surrounded by an asymmetric vitiligoid halo. [Copyright: ©2018 Lambertini et al.]

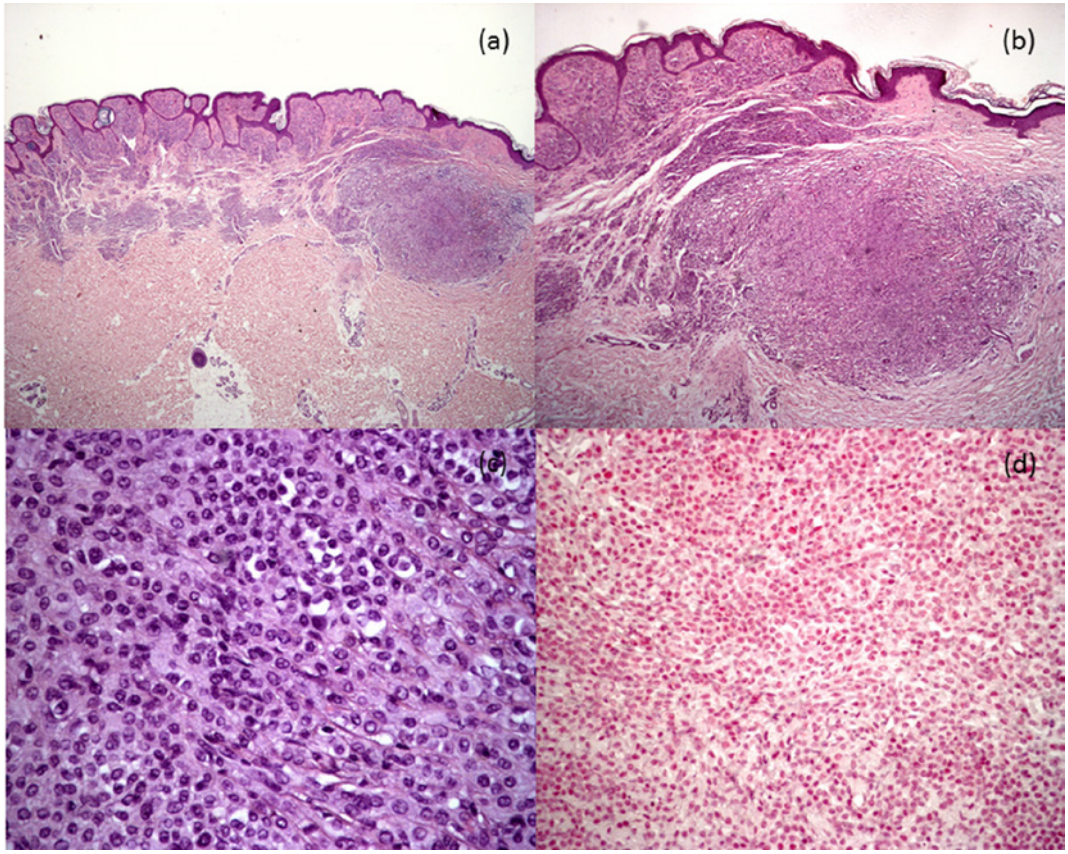

Figure 2. Histological findings. (a) Junctional and dermal melanocytic congenital nevus with a dermal nodule (hematoxylin-eosin, 4x). (b) Dermal nodule with nests of typical epithelioid cells. The periphery of the nodule showed gradual loss of cell density and blended with the surrounding nevus (hematoxylin-eosin 10x). (c) High power view. Large melanocytes with minimal pleomorphism and without necrosis (hematoxylin-eosin 40x). (d) BAP-1 immunostaining. Strong and diffuse nuclear staining in the proliferative nodule. [Copyright: ©2018 Lambertini et al.]

Answer

Proliferative nodule in a small congenital melanocytic nevus

Explanation

Proliferative nodules (PNs) are rare melanocytic neoplasms arising in congenital melanocytic nevi (CMNs) that usually have a very good prognosis but can mimic malignancy in the rapid onset and clinical features [1]. Nevertheless, histological analysis can be difficult and every new proliferation arising on a CMN poses a challenge to determine the differential diagnosis from a malignant melanoma [2-6].

Except for one case in a 17-year-old girl, PN proliferation has been described exclusively in newborns or during childhood [2-3]. Despite the worrisome clinical morphology, characterized by a nodular or papular consistency that slowly regresses, the majority of these neoplasms are benign, confirming the extreme rarity of melanoma in infancy [2-6].

Sometimes immunohistochemical analyses can be helpful, using the melanocytic markers S-100, HMB-45 or Melan A and other antibodies against Ki-67 and p-16 [7]. Immunohistochemical analysis was also performed in order to evaluate the loss of BAP-1 expression [8-10].

In our case, Ki-67 was moderately high in the PN and negative in the remaining congenital nevus, while p-16 was negative in the nodule, but positive in the remainder of the nevus. The presence of melanocyte maturation with symmetrical architecture and the presence of p-16 in the lower part surrounding the nodule did not support the criteria for malignant melanoma. BAP-1 immunohistochemistry revealed BAP-1 nuclear staining in the proliferative nodule, supporting the benign nature of the melanocytic proliferation [8-10].

Our case is remarkable because of the patient's age and the unusual clinical and dermoscopic features, characterized by an incomplete whitish halo corresponding to the focal scar and the reddish blotch on its top corresponding to the proliferative nodule on the left. It is possible, however, that her lesion had started primarily as a nodule, and the sudden occurrence of the depigmentation prompted the patient to seek medical advice.

In conclusion, PNs are typically found in childhood but may develop in much older individuals, and every rapid change on a CMN should always lead to surgical excision.

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