

In situ malignant melanoma on nevus spilus in an elderly patient

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Abstract

Nevus spilus is the term usually given to a pigmented skin lesion, congenital or acquired, that may occur anywhere on the body, consisting of a large light tan patch with numerous superimposed darker scattered maculae or papulae that are flat or slightly raised. For a long time, nevus spilus was believed to be a benign lesion. However, in 1957 Perkinson reported a melanoma appearing on nevus spilus for the first time. Since then other reports about melanomas developing on nevus spilus have been published, sometimes with a fatal outcome. We describe the case of an 80-year-old male patient with a congenital nevus just above his left knee. The lesion had remained unchanged over time, but some months before his checkup the patient noticed a darker area in the lesion that had continued to enlarge. The lesion was removed and histological examination revealed an in situ malignant melanoma. Although nevus spilus is not normally considered a precursor of melanoma, the potentiality of malignant transformation requires regular monitoring, and careful checkups are recommended and justified.

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Introduction

The term *nevus spilus* (NS) refers to hyper-pigmented patches of various sizes and sites, congenital or acquired, characterized by multiple darker brown speckled maculae, flat or raised, inside.

NS is a relatively common condition whose prevalence varies from 0.2 to 2.3%. As a congenital nevus, NS can be classified into three distinct clinical types (1, 2); small if it is less than 1.5 cm, medium if it varies from 1.5 to 19.9 cm, and large if its diameter exceeds 20 cm. There also exists segmental NS, better known as zoniform or zosteriform NS. NS can be found anywhere on the body, but the most common location is on the chest and upper limbs, and often the arrangement of NS follows the lines of Blaschko (3). Two distinct types of NS have been described, in the form of macular versus papular NS, each one sometimes related to a specific syndrome (4). The speckles are often maculae rather than papulae and develop during a period of months to years, with the number of elements ranging from eight to 10, and in some cases up to 30 elements, generally in relation to the size of NS. They usually range from 1 to 3 mm, but there can be a wide variety of size. The background of NS shows the histological features of a lentigo simplex or a café au lait macule, whereas the speckles show the histological features of a variety of nevi: junctional, compound, intradermal, dysplastic, Spitz, blue, neurotized nevus, or ink spot lentigo (3). Although as a rule NS is not considered a precursor of melanoma, it must be monitored because it may turn into a malignant melanoma.

We describe an in situ malignant melanoma in an 80-year-old male that developed on a congenital NS.

Case report

In an 80-year-old male patient, a newly formed irregular colored patch appeared on the right thigh inside a pre-existing speckled NS and was referred as fast-growing. The NS had been present at

birth and had not undergone any significant changes up to then. The patient's past medical history revealed the removal of a basal cell carcinoma and a malignant melanoma on his right leg 10 years earlier.



Figure 1 | Nevus spilus and melanoma. The examination shows the newly formed, tan to reddish-brown area arising on the congenital nevus spilus. The area is irregular and larger than the other speckles.

The clinical examination highlighted the presence of an oval-shaped patch, roughly 7 cm in diameter, pigmented, irregularly speckled with multiple random spread of flat maculae, darker than the patch itself (Fig. 2). More or less in the centre of the area there was a flat irregular 7 mm lesion, darker than the other maculae (Fig. 1).

The newly formed lesion was surgically removed and histopathologically showed to be “an in situ malignant melanoma” (Fig. 3a,b,c). Thereafter the patient was referred to a plastic surgeon for the complete removal of the remaining NS.

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Figure 2 | The complete feature of nevus spilus after biopsy of the in situ malignant melanoma. The oval patch is irregularly speckled with multiple random spread of flat maculae darker than the patch itself.

Discussion

For a long time NS was believed to be a benign lesion and, even if the development of melanoma on NS is a rare event, its possible malignant change has been widely reported. In 1957 Perkinson (5) described for the first time a melanoma appearing on NS in a patient with neurofibromatosis. Since then, 35 or more cases of melanoma developing on NS have been published, sometimes with a fatal outcome (6–11).

Reviewing all of the literature published, melanoma on NS usually seems to arise in Caucasian or black patients, more frequently on the trunk than the limbs, with a female-male ratio of 56:44 (2); the average age of patients is 49, range 17–79 years. Melanoma on NS is usually only one neoplasm, but sometimes there can be more (11).

The melanoma most often found is superficial spreading melanoma (68%), followed by nodular melanoma (16%), and only in a few cases in situ melanoma. Abecassis et al. (10) reported that in 52% of melanoma cases the NS was already present at birth, in 33% it was acquired during infancy, and in 14% it appeared later. In 60% of cases, melanoma was generated in small- to medium-sized NS with an average of 7.4 cm, in 24% of cases in zosteriform NS, and in the remaining 16% in giant NS. Given the scant number of giant NS, this confirms the higher progressive risk of larger NS (8). The presence of a dysplastic nevus on a NS is quite frequent (32% of cases), but its relevance is unclear.

From the studies published, the malignant potential appears to vary from 0.13% to 0.2% and seems to be higher in NS than congenital melanocytic nevi (12). Worthy of note is the fact that the macular NS can evolve into a melanoma more frequently than papular NS (4, 13).

Based on these general data, it is not possible to predict which patients may develop a melanoma on a NS; however, some important conclusions can be drawn. The risk factor of malignant transformation seems to increase when the lesion is congenital or acquired in infancy, when its size is ≥ 4 cm, and when the type of NS is macular rather than papular, taking into account the importance of the “nestling” cells in the hyperpigmented patch.

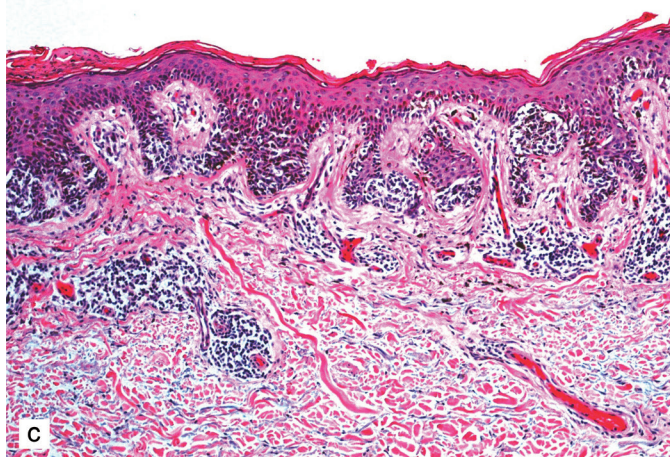
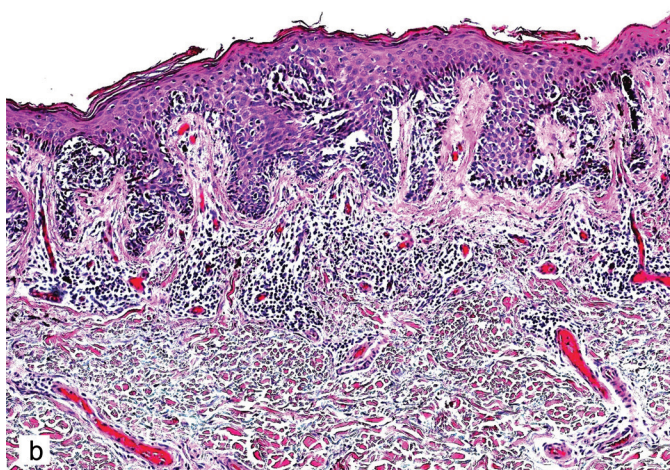
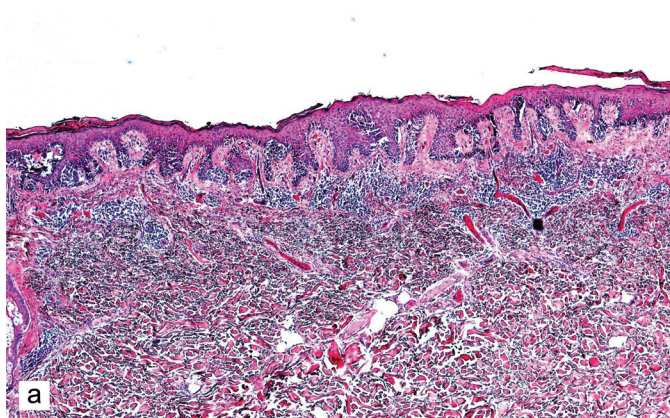


Figure 3a, b, c | Hematoxylin-eosin stain. Histopathological examination shows the characteristics of an in situ malignant melanoma, with the marked proliferation of atypical melanocytes along the dermal-epidermal junction, melanophage streams, and superficial inflammatory infiltration of the dermis. The raised melanocytes are inside the epidermis and do not create regular “nests”.

In the case of a positive histological test, complete excision of the entire lesion is necessary, size permitting, in order to eliminate the “faulty background” and to search for any multifocal melanomas. However, it is not justified to systematically remove all lesions, bearing in mind the low incidence of NS degeneration, unless the monitoring of the NS is problematic either because of its body site or the patient’s compliance. Finally, NS requires careful and closely individualized monitoring by dermatologists, and patients and relatives should be restricted to performing self-examination to detect early changes that may be the starting phase of turning malignant. Regular monitoring must also continue in elderly patients; a biopsy is recommended if a change is noticed in order to evaluate the his-

tological features and decide about excision.

Despite the wide range of publications, the link between NS and

melanoma still needs to be clarified. It is hoped that in the future targeted guidelines will be available for proper management of NS.

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