

Melanoma arising in a giant nevus spilus maculosus

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ABSTRACT

Melanoma arising in a nevus spilus is rare. There are two distinct types of nevus spilus characterized by macular or papular speckles, respectively. We report the case of a melanoma that arose in association with a giant nevus spilus maculosus.

Case report

A 78-year-old Caucasian woman presented with an ulcerated nodule in the right pretibial region (Figure 1). The nodule measured 1 cm in diameter and was surrounded by an erythema. It had grown in a large pre-existing, congenital pigmented lesion that, according to the patient, existed since birth (Figure 2). The gigantic nevus was flat and speckled. It extended over the right leg and the right side of the abdomen and the lower back, skipping the buttock and the back of the thigh, and crossed the midline (Figure 3). Close up it consisted of multiple dark-brown macules on a light brown, café-au-lait-like background (Figure 4). A punch biopsy of the nevus showed elongation of rete ridges and hyperpigmentation of basal keratinocytes but no substantial proliferation of melanocytes (Figure 5). Histopathology of the nodule showed a nodular melanoma with an invasion thickness of 4.5 mm (Figure 6). On the basis of the clinical and pathologic findings we made the diagnosis of a melanoma arising in a giant nevus spilus maculosus.



Figure 1. Close-up of eroded nodule with surrounding erythema.



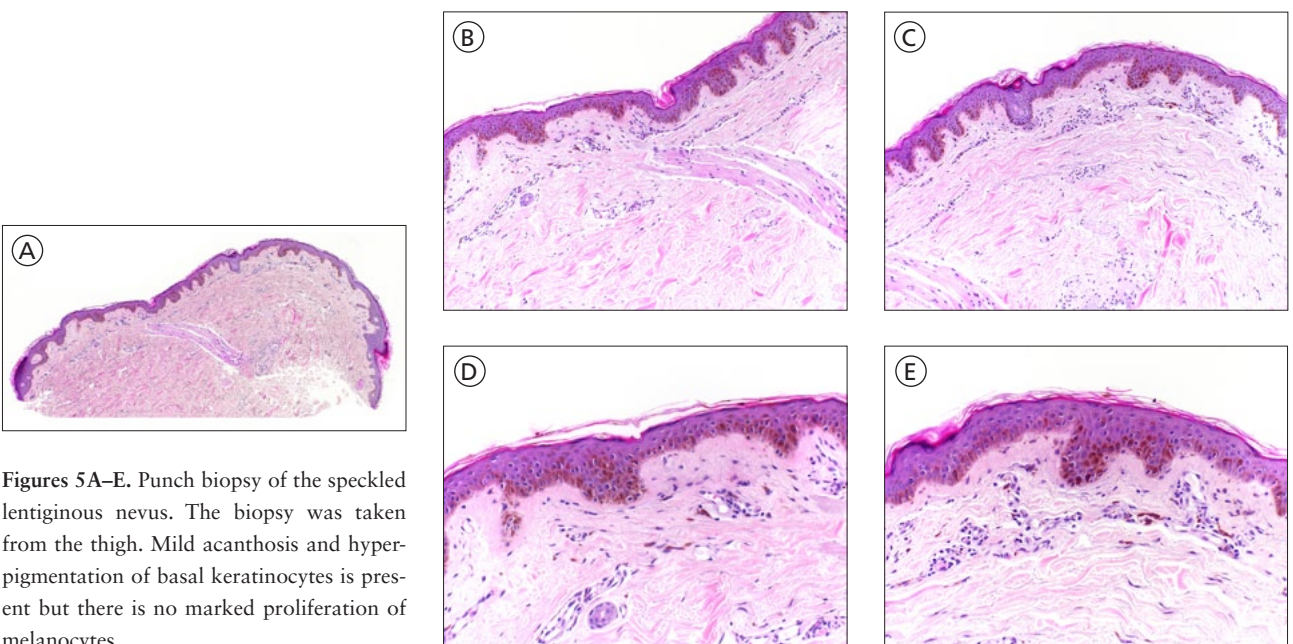
Figure 2. The nodule on the right leg appeared in association with a speckled lentiginous nevus (nevus spilus).



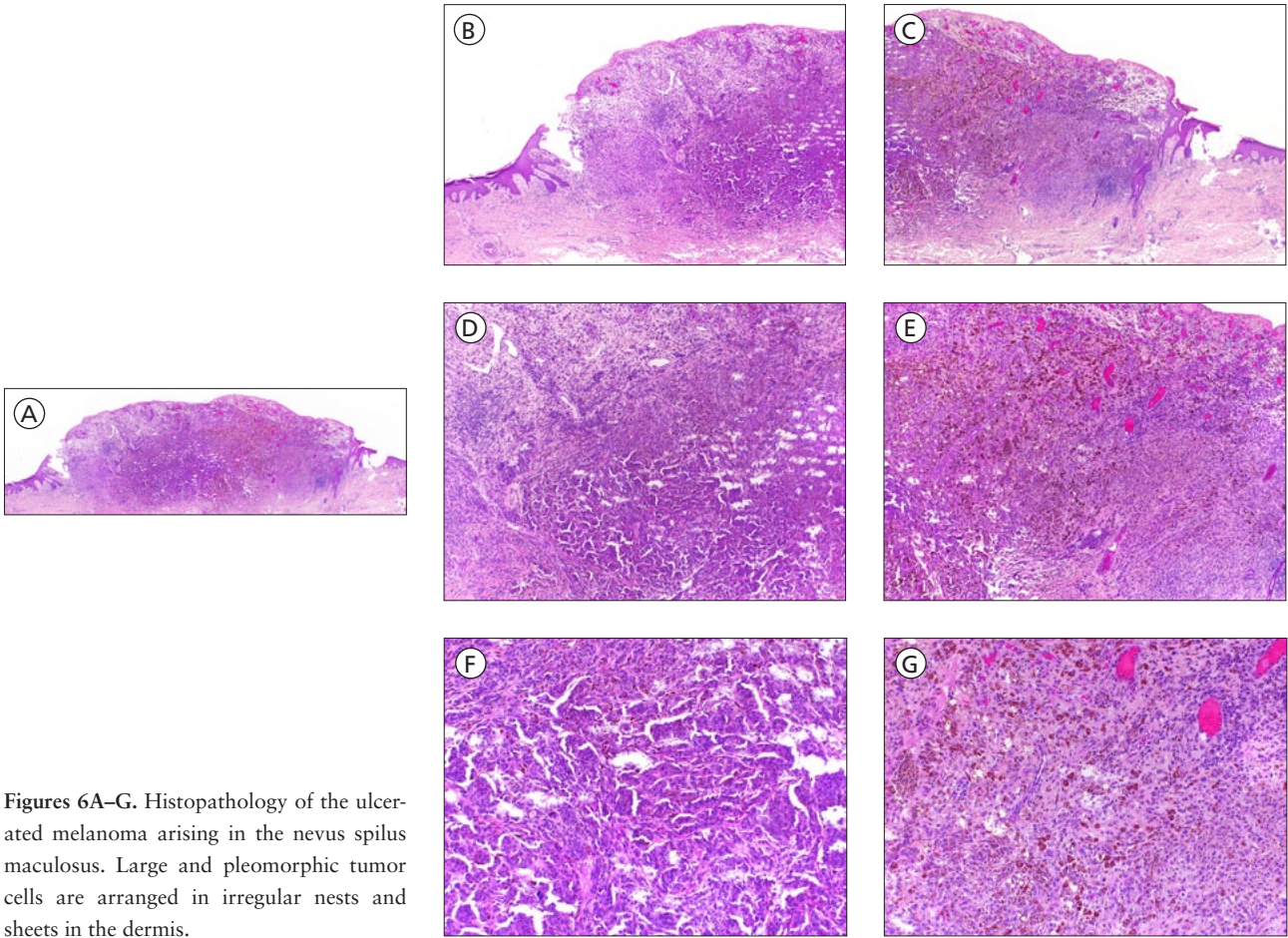
Figure 3. Extension of the nevus spilus that crosses the midline.



Figure 4. Close-up of nevus spilus maculosus.



Figures 5A–E. Punch biopsy of the speckled lentiginous nevus. The biopsy was taken from the thigh. Mild acanthosis and hyperpigmentation of basal keratinocytes is present but there is no marked proliferation of melanocytes.



Figures 6A–G. Histopathology of the ulcerated melanoma arising in the nevus spilus maculosus. Large and pleomorphic tumor cells are arranged in irregular nests and sheets in the dermis.

Discussion

Malignant melanoma arising in association with a nevus spilus, also referred to as speckled lentiginous nevus, is rare [1,2]. In German, nevus spilus is also named “Kiebitzei-Naevus,” referring to the peculiar pattern of the nevus that resembles a lapwing’s egg (Figure 7).

According to Happle there are two distinct types of nevus spilus [3,4]. Nevus spilus maculosus is flat and characterized by dark brown spots on a light brown background. In this type of nevus spilus the background pigmentation histopathologically corresponds to hyperpigmentation of basal keratinocytes, and the dark spots are believed to be typified by junctional melanocytes arranged in nests. In the presented case a punch biopsy of the nevus did not show a proliferation of melanocytes, but the biopsy was relatively small and sampling error cannot be excluded.

Papular nevus spilus, on the other hand, is characterized by dark brown papules or nodules on a light-brown background, although small dark spots may also be present [3]. Histopathologically, this type of nevus spilus shows the pattern of a superficial and deep congenital nevus with the presence of melanocytes in the papillary and reticular dermis. The malignant potential of the papular type appears to

be lower than that of macular type of nevus spilus [3]. It is possible that the two different types of nevus spilus originate from different mutations. This would also explain the dissimilar malignant potential of either type of nevus [4].

As a possible differential diagnosis we also considered a partial unilateral lentiginosis (PUL), which seems to be a



Figure 7. Four lapwing’s eggs [German: Kiebitz Eier]. Picture courtesy of Dr. Ernst Bauernfeind, Head Curator of the Bird Collection, Museum of Natural History, Vienna, Austria).

mosaic manifestation of type 1 neurofibromatosis [5]. This rare condition was excluded on the basis of clinical findings because in PUL the background is normal and not hyperpigmented and the body-midline is not crossed. In addition, our patient showed no signs of neurofibroma or café-au-lait macules [6,7]. Phacomatosis spilorosea, a subtype of phacomatosis pigmentovascularis, could also be excluded because of the absence of telangiectasias [8]. We excluded the speckled lentiginous nevus syndrome that is characterized by a speckled lentiginous nevus of the papular type and ipsilateral neurological abnormalities such as hyperhidrosis, muscular weakness and dysesthesia because such abnormalities were not present in our patient [9].

This case of a melanoma arising in a nevus spilus maculosus suggests that this type of nevus should be regularly observed by a dermatologist and patients should be instructed to perform self-examinations to detect changes as early as possible.

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