

Nevoid Melanoma in the Forearm

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Abstract

Nevoid melanoma is a very rare histological subtype of vertical growth phase melanoma. Histologically, it mimics benign nevus and thus may lead to an erroneous diagnosis. We report a case of nevoid melanoma arising in a 53-year-old American woman. High index of suspicion and evaluation of cytologic atypia with ancillary tests may help in establishing the diagnosis.

Key Words : Melanoma, Skin

Introduction

Differentiation of benign and malignant melanocytic lesion is a challenging issue both for clinicians and pathologists. Among melanomas, desmoplastic melanoma, acral-lentiginous melanoma and nevoid melanoma are most commonly misdiagnosed [1].

Well-known clinical features for the diagnosis of malignant melanoma include asymmetry, irregular border, uneven color and size larger than 6mm. These features are usually applied to plaques and patches of radial growth phase

melanoma [2]. Histologic diagnosis of melanoma depends on the combination features of poor circumscription, size, asymmetry, pagetoid spread, lack of maturation, cytologic atypia and mitotic figures [3].

Nevoid melanoma is a very rare type of melanoma with less than 1% of melanomas [4,5]. Especially with low-power microscopic examination, misdiagnosis as benign nevus is frequently rendered [6,7]. It resembles a nevus in that it shows symmetrical growth architecture, variable maturation and no prominent intraepidermal component. It manifests sharp

lateral circumscription. Junctional activity is uncommon but can be observed [4]. However, at high-power examination, the tumor cell shows mitosis in deep dermis and cytologic atypia with prominent nucleoli at base. Nevoid melanomas are usually devoid of malignant features such as a pushing border, high mitotic activity and striking pleomorphism [4]. Diagnosis of nevoid melanoma depends on high index of suspicion and careful evaluation of cytologic features.

We report a case of nevoid melanoma in the right forearm of a 53-year-old white woman. No case has been reported in Koreamed.

Case Report

A 53-year-old white woman presented with a polypoid mass of several years of history on the right forearm (Fig. 1). The mass easily bled and appeared to be growing recently. She had previous history of chronic pancreatitis and was otherwise unremarkable. The clinical diagnosis for the mass was hemangioma. A punch biopsy was obtained and a diagnosis of nevoid melanoma was suggested. The patient underwent wide excision with no additional treatment. With 6 years of follow-up, the patient showed no evidence of recurrence.

Grossly, the mass was 1.0 cm in greatest dimension, showing a relatively well-defined reddish pink, round and polypoid appearance. Microscopically, the mass consisted of dense proliferation of polygonal to oval epithelioid cells, involving dermis. The lesion was relatively well-defined and symmetrical (Fig. 2). The tumor cells were arranged in sheet and occasionally in nests. Melanin pigments were observed within the cytoplasm of tumor cells. They showed cytologic atypia with hyperchromatic nuclei and prominent



Fig. 1. A finger-tip sized protruding reddish mass in the right forearm.

nucleoli (Fig. 3). Mitotic figures were observed throughout the tumor up to 10/10HPFs. There is no maturation with depth. Pagetoid spread in epidermis was not observed. Mild to moderate lymphocytic infiltration was associated. The tumor cells were positive for HMB45 and S-100 immunostainings. The Ki-67 proliferation index (PI) was about 10% throughout the lesion. Immunostaining for cyclin D-1 showed diffuse positivity both in superficial and deep components of tumor (Fig. 3). With these histologic features, the diagnosis of nevoid melanoma was rendered.

Discussion

Nevoid melanoma is a rare histological subtype of vertical growth phase melanoma. It was first proposed by Schmoeckel *et al* and shows deceptive morphologic features reminiscent of a benign melanocytic nevus. It is one of the most commonly misdiagnosed melanoma and carries

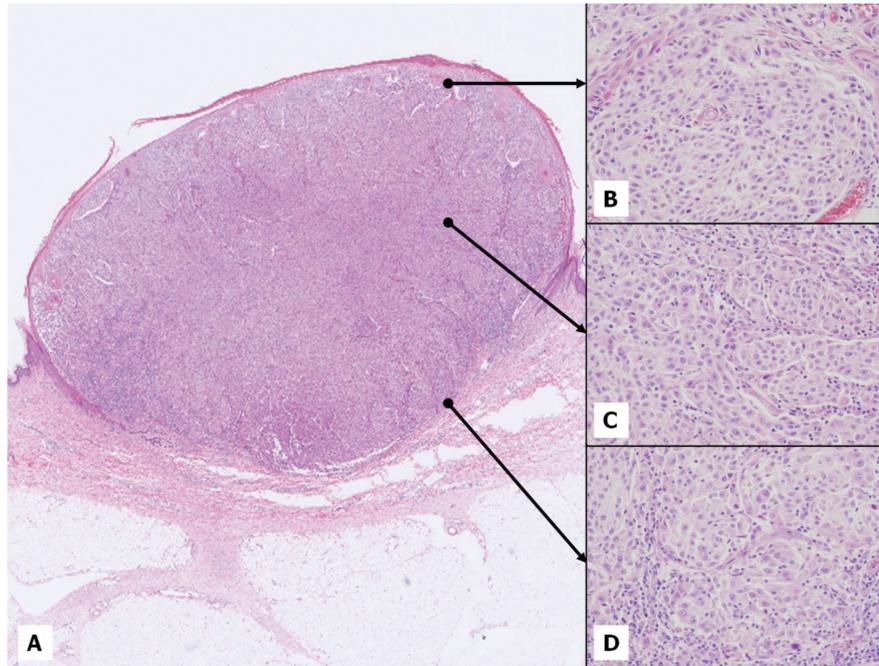


Fig. 2. Low magnification view of the mass shows symmetric and well-circumscribed growth (A). The superficial (B), middle (C) and base (D) parts show similar pattern without maturation.

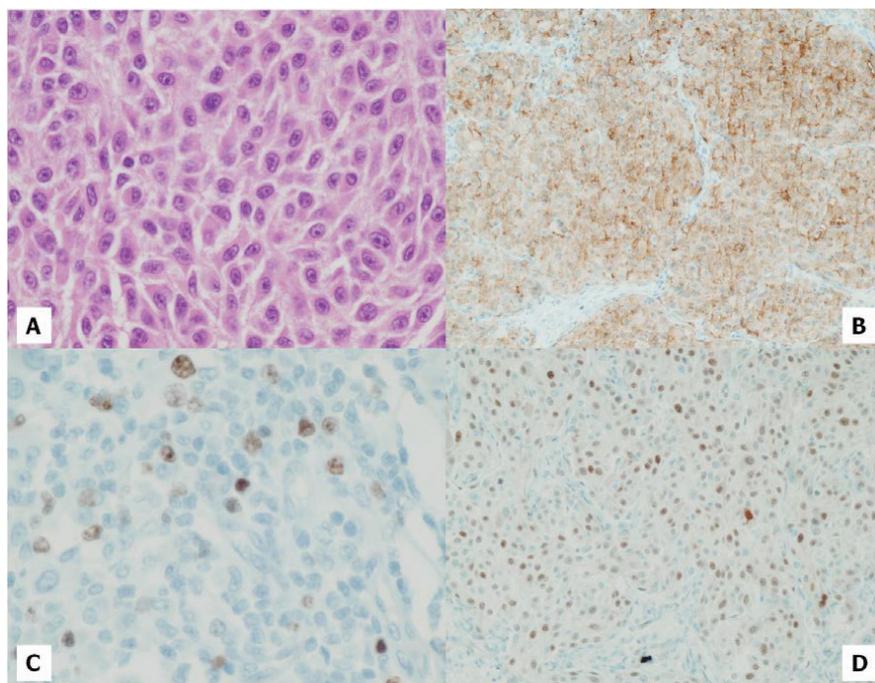


Fig. 3. The tumor cells are relatively monomorphic but show cytologic atypia with prominent nucleoli (A). The immunohistochemical staining for S-100 shows positivity (B). The Ki-67 proliferation index is about 10 % (C). The immunohistochemical staining for cyclin D-1 shows diffuse positivity (D).

significant medicolegal implications [1,8,9]. The incidence is apparently very low but no reliable and unbiased data have been available. The demographic characteristics are controversial in terms of sexual predilection and sites [4,10]. Clinically, there is no particular distinguishing features, most lesions being clinically described as verrucous to dome-shaped variably pigmented nevi or non-specific papules or nodules. Our case presented with a protruding finger tip-sized nodule with easy bleeding and was initially considered as hemangioma. The prognosis has not been clearly defined. A follow-up study indicated a recurrence rate of 50%, a metastatic rate of 25~50% and a mortality rate of at least 25% [9]. More studies with large number of case are required for further clarification.

Histologically, the diagnosis of nevoid melanoma is very challenging in that important criteria for the diagnosis of melanoma are usually missing. Asymmetrical growth, poor circumscription and pagetoid melanocytosis are absent and maturation may be present. The present case also showed symmetrical growth, well circumscription and no pagetoid spread. Barnhill *et al.* suggested several clues to the diagnosis of nevoid melanoma, which include dermal mitoses, sheet-like appearance, monomorphous appearance of melanocytes, subtle but definite cytologic atypia, lack of conventional maturation and presence of irregular infiltrating features at the base and angiotropism [10]. Nevoid melanoma can be distinguished from benign melanocytic lesion with a high index of suspicion and a careful evaluation of the architecture and cytologic features. Immunohistochemical staining may be of help in diagnostically challenging case. High Ki-67 PI and nuclear staining of cyclin D1 throughout the depth of the tumor support the diagnosis of nevoid melanoma [9]. The present case showed definite

cytologic atypia and atypical mitosis without maturation as well as high Ki-67 PI and diffuse immunopositivity for cyclin D1 throughout the lesion. If diagnostic difficulties persist between benign nevus and melanoma, it is recommended to seek second opinion. Variable molecular tests such as comparative genomic hybridization or fluorescence in situ hybridization using probes targeting 6p25, 6q23, Cep 6 and 11q13 may assist in establishing the diagnosis [11-13]. However, the clinical application of these studies needs further independent validations [12]. To avoid misdiagnosis, excisional biopsy rather than punch or shave biopsy and clinicopathologic correlation are recommended [1].

Wong *et al.* described two architectural patterns of nevoid melanoma, dome-shaped and verrucoid, respectively [7]. The dome-shaped tumors showed smooth epidermal surface and proliferation of epithelioid cells with an inconspicuous intraepidermal component as in the present case. The verrucoid lesions were characterized by broad, exophytic tumors with a verrucous epidermal surface. The differential diagnosis of nevoid melanoma among malignant melanoma includes minimal-deviation melanoma, nodular melanoma, metastatic melanoma and melanoma arising in a dermal nevus [4,10].

In conclusion, nevoid melanoma mimics the benign nevus and carries significant medicolegal implications. High index of suspicion and evaluation of cytologic atypia with ancillary tests may help in establishing the diagnosis.

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