Clinical Letter

Primary cutaneous angiomatoid melanoma

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Dear Editors,

A 63-year-old woman with no relevant medical history presented to our hospital with changes in the morphology and color of a chronic cutaneous lesion. Two weeks prior to consultation, the lesion had bled profusely, subsequently decreasing significantly in size. Dermatological examination revealed a purplish tumor (1.5 cm in diameter) in the right scapular region, arising from a slightly raised brown papule. There were signs of recent bleeding and some crusting. Dermatoscopy showed a violaceous-black homogeneous pattern and a blue white veil, with white fibrous tracts and irregular vessels inside. Beneath, a brown melanocytic lesion with a regular cobblestone pattern was observed (Figure 1). The remainder of the clinical exam was unremarkable.

An excisional biopsy was performed. Histopathological examination revealed a well-nested melanocytic lesion with an extensive intradermal component, progressive in-depth maturation, and no signs of atypia. A nodular tumor whose largest diameter was 0.7 cm grew adjacent to the aforementioned nevus; it was composed of medium-sized cells with marked pleomorphism, clear nuclei, and prominent nucleoli with frequent mitoses. There was a lack of maturation throughout the neoplasm. Atypical melanocytic cells arranged in a lentiginous pattern without nest formation were present along the basal layer. There were scattered tubular structures resembling vascular channels containing numerous erythrocytes. These channels were lined by neoplastic cells that were negative for endothelial markers D2-40 and CD-31. Further immunohistochemical staining showed consistent positivity for melanocytic markers (S-100, SOX-9, and HMB-45) and negativity for cytokeratins A1–A3 (Figure 2).

Given the clinical and histological findings, a diagnosis of primary angiomatoid melanoma (Clark level III, Breslow thickness 5.2 mm, with evidence of ulceration between 3–5 mm) was established. Whole-body positron emission tomography-computed tomography (PET-CT) was performed, revealing metastatic disease in the lungs, liver, and celiac lymph nodes. Genetic testing for both V600E and V600K mutations of the *BRAF* gene was performed using real-time PCR (THxID-BRAF AMP, bioMérieux, France), revealing a V600E mutation. Following the diagnosis, the patient was referred to the oncology department and started on vemurafenib (960 mg BID). After six months of follow-up, there was no evidence of disease progression.

Cutaneous melanoma is characterized by clinical and histopathological variants, the latter including an increasing number of different designations [1] (including nevoid, follicular, animal, spitzoid). Rarely reported in the literature, angiomatoid pattern is an extremely uncommon histological melanoma variant [2, 3]. It was first described by Adler et al. in 1997 in a patient with cutaneous metastatic melanoma of unknown origin [4]. As in our case, the lesion on the forehead showed clusters of HMB-45 and S-100-positive cells forming vascular channels [4]. Since then, four more cases with similar histopathology have been published [2, 3]. A vessel-like morphology with pseudovascular channels has also been described in other melanocytic neoplasms [5, 6]; here, due to the lack of tissue resistance to traction, vascular spaces may be formed as a result of the biopsy procedure. In addition, these vessel-like formations can be induced by tumor cell vasculogenic mimicry, a phenomenon observed in a

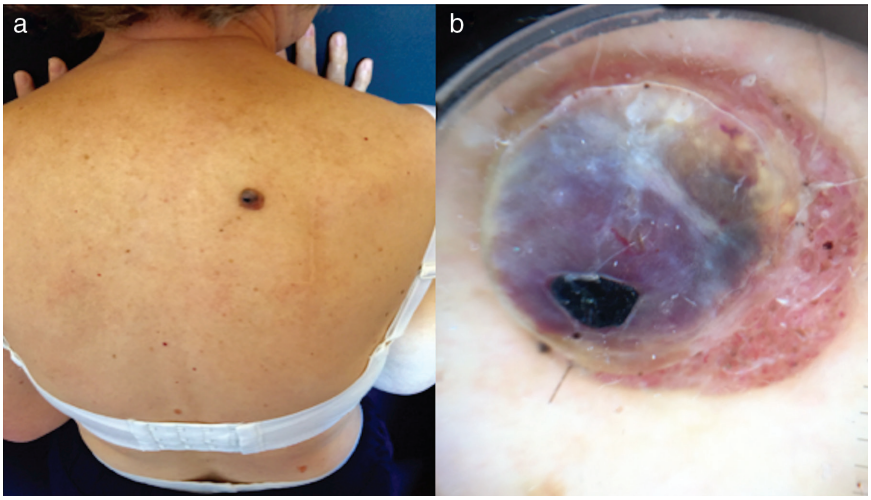

Figure 1 Clinical presentation and dermatoscopy of angiomatoid melanoma. Clinical picture. Violaceous nodule on top of a brown papule with a nevoid appearance (a). Dermatoscopy showing a brownish melanocytic lesion with pink areas and a global cobblestone pattern. Adjacent to this nevus, there is a homogeneously purple nodular formation with lacunae-like structures separated by white fibrous linear septa and a central blue-white veil. Note the markedly irregular vessel within one lacunar structure (b).

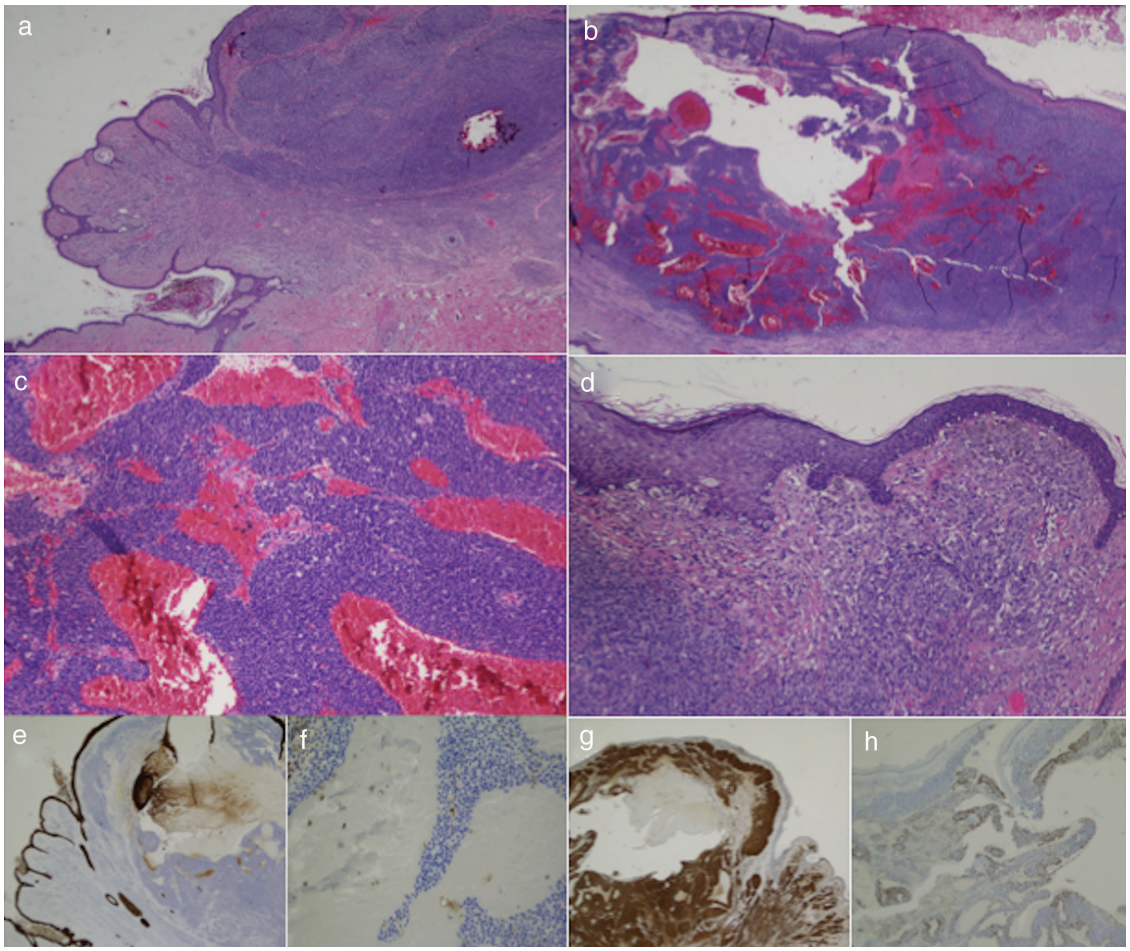

Figure 2 Histopathology. There are two different neoplasms. On the left side, there is an intradermal nevus with regular melanocytes with progressive dispersion arranged throughout the dermis. On the upper right side, there is a strongly cellular nodule deeply invading the dermis (hematoxylin-eosin stain [HE], original magnification x 4) (a). Picture of the aforementioned nodule occupied by large channels full of erythrocytes (HE x 4) (b). Close-up of the pseudovascular spaces. They are surrounded by medium-sized pleomorphic cells. There is no endothelial lining (HE x 20) (c). Photograph of the epidermis and papillary dermis. There are numerous atypical melanocytic cells that invade the basal layer, showing a lentiginous pattern (HE x 20) (d). Immunohistochemistry. Both the intradermal nevus and the melanoma are negative for cytokeratins A1–A3 (e); the pseudovascular channels are negative for the lymphatic endothelial marker CD-31 (f). The melanoma and the adjacent nevus are distinctly positive for S-100 (g) and HMB-45, with a focal pattern in the latter (h).

large number of aggressive tumors in which cancer cells create a pseudovascular network within the tumor core due to genetic dedifferentiation and expression of mesenchymal genes. Such a network may provide an escape route for metastasis [7]. Vasculogenic mimicry has previously been described in a uveal melanoma with an angiomatoid pattern [8].

We are the first to report that angiomatoid melanoma may present with a dermatoscopic picture resembling that of vascular neoplasms such as pyogenic granuloma and senile angiomas, showing a homogeneous pattern, lacunae, and white fibrous linear septa [9, 10]. However, in the present case, there were highly suspicious signs of malignancy such as irregular vessels and a blue-white veil. The prominent structureless bluish color with scattered clods was also highly suspicious of melanoma [11]. The melanoma in our patient grew adjacent to an intradermal nevus, and there have been some reports of intradermal melanoma associated with an intradermal nevus [12]. Based on the histopathological information, we consider this a coincidental finding, since that particular melanoma had a prominent intraepidermal and junctional component. In addition, there was a sharp demarcation between the two neoplasms.

As in the present case, angiomatoid melanoma usually shows an aggressive behavior. While it tends to exhibit metastatic histological features [3, 4], there are at least two reports of primary angiomatoid melanoma, as in our patient [2, 3].

Conflict of interest

None.

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