

# **RESEARCH ARTICLE**

### ATYPICAL MELANOMA MIMICKING A VASCULAR TUMOR : CLINICAL AND DERMOSCOPIC FINDINGS

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## Manuscript Info

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#### Abstract

**Introduction:** Completely amelanotic melanomas are rare and therefore often misdiagnosed. Analysis of dermoscopic features of the vascular pattern appears to be a reliable method to suggest these particular diagnosis.

**Case Presentation:** we report a case of an atypical primary cutaneous melanoma of plantar localization which clinically mimicked an angiomatous tumor. Clinical examination showed an angiomatous lesion of 10 cm long axis, located on the lateral part of the left sole. The two adjacent nodules measured 3 cm each, one was angiomatous and the other erythematous. There were also multiple left inguinal adenopathies. Dermoscopic evaluation of the lesion revealed the lack of a pigmented network and a rich vascular bundle made of polymorphous linear vessels, dotted vessels, white lines and \"milky red\" areas. The patient underwent excision of an angiomatous nodule. The definitive diagnosis was provided by immunohistochemistry (tumor cells expressed S100 and Melan A). Brain and thoracic-abdominal-pelvic CT scan showed bilateral nodular lung involvement and a segment VI liver lesion. We concluded to the diagnosis of a primary amelanotic metastatic melanoma stage 4.

**Clinical Discussion:** The term amelanotic refers to tumors that have no pigmentation on visual and dermoscopic inspection. Amelanotic and hypomelanotic melanomas are rare. They could mimic many benign and malignant tumors. There are three forms of AM. The papulonodular form represents 58% and could be misdiagnosed as a vascular tumor, which was the case of our patient. The contribution of dermoscopy is valuable in the clinical orientation in case of amelanotic melanoma. There is a correlation between the stage of the melanoma and the vascular pattern(s) observed as well as their distribution. The prognosis of amelanotic melanoma is characterized with a lower survival rate and a high risk of recurrence. The treatment of amelanotic melanoma is similar to the pigmented counterpart.

**Conclusion:** In order to avoid therapeutic delay, melanoma should be considered whenever there is a suspicious plantar lesion even in the absence of pigmentation. Dermoscopy is a non-invasive technique that could help in the diagnosis.

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#### Introduction:-

Among all skin tumors, melanoma has the worst prognosis [1]. Clinically, it is recognized by its blackish pigmented color. However, it is possible to observe a poorly pigmented or non-pigmented lesion, simulating a variety of non-melanocytic tumors [2]. We report a case of an atypical primary cutaneous melanoma presenting as an angiomatous tumor of plantar localization. The work has been reported in line with the SCARE 2020 criteria [3].

#### **Case Report**

A 53-year-old man presented with an enlarging plantar budding lesion on the left foot of more than 2 years duration, that has progressively became painful. Two nodules appeared nearby 18 months later. On clinical examination, the lesion was angiomatous, 10 cm long axis, located on the lateral part of the sole of the left foot (fig.1). The two adjacent nodules measured 3 cm each, one was angiomatous and the other erythematous (fig.2). There were also multiple left inguinal adenopathies.

Dermoscopy showed a rich vascular bundle made of polymorphous linear vessels, dotted vessels, white lines and "milky red" areas (fig.3). There was no pigmentary network. An excision of the angiomatous nodule was performed. Histology revealed a malignant melanocytic proliferation confirmed by immunohistochemistry (tumor cells expressed S100 and Melan A) with a Breslow score of 8 mm and a Clark and Mihm level IV (fig.4, 5). Mitoses were 2/mm<sup>2</sup>. MRI of the forefoot showed deep extension to the regional muscles and envelopment of adjacent vascular structures. Brain and thoracic-abdominal-pelvic CT scan showed bilateral nodular lung involvement and a segment VI liver lesion.

We concluded to the diagnosis of a primary amelanotic metastatic melanoma stage 4. Taking into account the extensive local invasion and the metastatic aspect, the decision of the multidisciplinary consultation meeting was to start palliative care in association with amputation of the foot. As the patient refused amputation, local radiotherapy was suggested.

#### **Discussion:-**

The term amelanotic refers to tumors that have no pigmentation on visual and dermoscopic inspection [1,4]. Amelanotic and hypomelanotic melanomas are rare. They represent 2 to 8% of all melanomas [2]. The average age is over 50 years and the sex ratio M/F varies from 0.5 to 4. The risk factors are phototype I, oculocutaneous albinism, presence of ephelides, absence of naevi on the back, photosensitivity and family history of amelanotic melanoma (AM) [5].

The mechanism of amelanotic melanoma is still unclear. It is actually controversial that AM consists of poorly differentiated or dedifferentiated tumor cells [6]. Instead, it appears that AM cells retain their ability to synthesize melanin via the expression of Tyrosinase and Microphthalmia associated Transcription Factor (MITF) [5]. On the other hand, there is a phenotypic transformation causing the decrease of expression of the specific enzymes of this synthesis suggesting that amelanosis or hypomelanosis is due to an insufficient quantity or activity of Tyrosinase [5,6].

Due to the lack of pigmentation, AM could mimic many benign and malignant tumors. There are three forms of AM. The papulonodular form represents 58% and could be misdiagnosed as a vascular tumor, which was the case of our patient in whom Kaposi's sarcoma and angiosarcoma were the first considered diagnoses. The other two forms are the erythematous macule with epidermal changes and the dermal plaque without epidermal changes [5].

The contribution of dermoscopy is valuable in the clinical orientation in case of amelanotic or hypomelanotic melanoma [7]. It improved the sensitivity (from 65 to 89%) and specificity (from 88 to 96%) of the diagnosis of AM compared to naked eye inspection [8]. This is possible through the analysis of the vascular bundle [9]. There is a correlation between the stage of the melanoma (macular, elevated, nodular lesion) and the vascular pattern(s) observed as well as their distribution. In the early stages, punctiform vessels with a homogeneous appearance and regular distribution are most often observed. When the tumors are thick, the vascular bundle is polymorphic, with longer, thicker, irregularly distributed vessels. 'Milky red' globules are more frequent [7]. In addition to the vascular

bundle, the most important dermoscopic criteria are ulceration, scar-like depigmentation, white streaks and pigmentary network borders [5].

The amelanotic variant is most commonly found in melanomas of subungual location (25%) and in desmoplastic melanomas (46-93%) [1,5]. Acrolentiginous melanomas are amelanotic in 17-27% [5]. In our patient, the plantar location and the acrolentiginous type on histology attribute originality to our case. AMs are associated with a higher Breslow index, Clark stage and independently with a higher mitotic index [10,11].

The prognosis of ADs is characterized with a lower survival rate and a high risk of recurrence. Moreover, they are diagnosed at advanced stages: Some authors suggest that this is due to diagnostic delay, others incriminate the intrinsic aggressiveness of these tumors [5]. The treatment of amelanotic melanoma is similar to the pigmented counterpart [1].

Figures



Figure 1:- Angiomatous erythematoviolaceous plantar lesion.



Figure 2:- Two adjacent plantar nodules.



Figure 3:- Dermoscopy findings : Polymorphous linear vessels, dotted vessels, white lines and "milky red" areas.



Figure 4:- Immunochemistry findings: tumors cells expressing S100 (\*20).



Figure 5:- Immunochemistry findings: tumors cells expressing Melan A (\*40).

### **Conclusion:-**

AM is a challenging disease because of its nonspecific clinical manifestations [4]. In order to avoid diagnostic and therapeutic delay, melanoma should be considered whenever there is a suspicious plantar lesion even in the absence of pigmentation, especially in Maghreb region. This aims to prevent heavy repercussions on the patient. Dermoscopy is a non-invasive technique that could help in the diagnosis [4].

#### **Patient Consent**

Written consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the edito-in-Chief of this journal on request.

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