Summary

Diffuse melanosis cutis (DMC) is a rare presentation of metastatic melanoma characterized by a progressive blue-grey discoloration of the skin and mucous membranes.

A 60-year-old female patient is consulted for diffuse brown-grey discoloration of the skin located on the chest and the left upper limb which appeared 2 months ago and had a progressive evolution. The personal history of the patient shows a malignant melanoma operated on 8 months ago and currently with skin and liver metastases.

DMC is the progressive discoloration of the skin and mucous membranes in metastatic malignant melanoma. The release of melanin from melanoma metastases and its phagocytosis by dermal histiocytes is currently recognized as the cause of DMC. DMC prognosis is poor.

Introduction

Diffuse melanosis cutis (DMC) is a rare manifestation of malignant melanoma characterized by pigmentation of skin and mucous membranes in the context of a metastatic melanoma. The original description of DMC is attributed to Wagner in 1864.

Clinical case

A 60-year-old female patient is consulted for a diffuse discoloration of the skin which appeared 2 months ago and had a progressive evolution. The personal history of the patient shows a malignant melanoma operated on 8 months ago with a 2.10 mm Breslow’s depth and a Clark level IV, located in the upper back region.

The dermatological examination reveals multiple, enlarged plaques of different sizes, of brown-grey colour located on the anterior (Fig. 1) and posterior (Fig. 2) thorax, the left shoulder and arm. The histopathological examination shows the presence in the dermis of melanin deposits intracytoplasmically, but also extracellularly in the connective tissue with predominantly perivascular disposition.

The patient has on the right shoulder a metastatic skin tumour formation (Fig. 3), but also liver metastases evidenced by magnetic resonance imaging. The patient’s urine was black.

Discussions

Diffuse melanosis cutis (DMC) is characterized by the appearance of a rapidly progressive cutaneous-mucosal pigmentation in the context of a metastatic mucosal melanoma. The colour of the pigmentation is variable, ranging from the
colour of slate to dark blue-black with a cephalocaudal progression. The pigmentation distribution is more pronounced on the photo-exposed areas, being more frequently met in the form of “splashes or drops” and less frequently in a diffused form as in the case presented by us. Melanuria or black urine is found in almost all patients with DMC. Usually the urine sample must be exposed for a long time to light and air to turn black. Melanuria may also occur in association with metastatic melanoma in the absence of DMC (2). Other clinical manifestations such as hair (3,4), serum (5) and peritoneal fluid6 blackening may occur in DMC as well, and melanin may also be present in expectoration5. Pigmentation of the mucous membranes5,7,8,9 as well as melanosis of the internal organs, including bone marrow (2,3,5,7,10,11,12) may also be encountered.

Histology highlights the growth of melanic pigment in the dermis both in histiocytes, and also as free pigment in dermal connective tissue, fibroblasts and in cellular debris, having a particularly perivascular disposition. In the epidermis there is only an increase of keratinocyte pigmentation. Electron microscopy shows changes identical to those found in optical microscopy(3).
The pathogenesis of DMC remains unclear. It was initially thought that its cause was melanoma cell migration into the dermis, this being responsible for melanin storage (13). The absence of tumour cells in the dermis has dismissed this theory. Another theory involves in the occurrence of DMC the melanocyte growth factors such as α-melanocyte stimulating hormone, endothelin-1 and hepatocyte growth factor that would activate melanogenesis, proliferation and spread of normal and malignant melanocytes (10), a theory denied by the absence of alteration of the number of melanocytes or pigmented keratinocytes. The release of melanin, its pre-cursors and melanosomes from melanoma metastases and their phagocytosis by dermal histiocytes is currently recognized as the cause of DMC(14).

The average duration between the diagnosis of melanoma and the occurrence of melanosis in the same patient is less than 1 year, and the survival from the time of onset of DMC is of approximately 4 months. In conclusion, DMC is a rare manifestation of metastatic malignant melanoma, having a poor prognosis.

Bibliography


Conflict of interest
NONE DECLARED

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