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Dermoscopic findings in an early malignant fibrous histiocytoma on the face

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ABSTRACT Malignant fibrous histiocytoma (MFH), currently classified as undifferentiated pleomorphic sarcoma, is the most frequent soft tissue sarcoma in adulthood, but it is not as common as a primary skin tumor. MFH affects mostly the thighs and trunk, head and neck is an infrequent presentation in adults. MFH is often diagnosed in advanced stages, with a tendency to local recurrence and systemic metastasis. Since tumor thickness and size are identified as major prognostic factors, early recognition becomes crucial to improve prognosis. We present a case of a cutaneous malignant fibrous histiocytoma located on the face in which dermoscopy was useful in clinical management and definition.

Introduction

Malignant fibrous histiocytoma (MFH), currently classified as undifferentiated pleomorphic sarcoma, is the most frequent soft tissue sarcoma in adulthood, but it is not as common as a primary skin tumor [1]. The age of presentation ranges between 50 and 70 years, two-thirds occur among men, and the Caucasian population is more commonly affected. MFH affects mostly the thighs and trunk. Infrequently, it presents in the head and neck in adults [2,3]. The lesions are usually diagnosed in advanced stages, and despite currently proposed therapies such as radiotherapy or chemotherapy, the patient's prognosis is usually poor with a tendency to local recurrence and systemic metastasis [1]. Early recognition is crucial to improve clinical outcome. Dermoscopy has shown to be useful in the assessment of both melanoma and non-melanoma skin tumors [4].

Case Report

A 75-year-old male with history of multiple basal cell carcinomas and actinic keratosis presented for a biannual routine skin examination. In the left cheek a hypopigmented lesion was detected. It was discretely erythematous, with poorly defined limit, and increased consistency on palpation (Figure 1). The lesion was not present at the time of his previous visit and the patient was not aware of the lesion and did not know the evolution. No previous history of radiotherapy

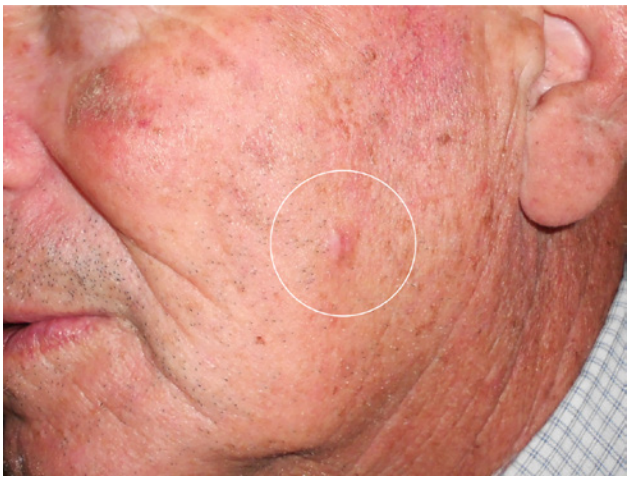

Figure 1. Clinical image. Hypopigmented lesion with discrete erythema and poorly defined limits. [Copyright: ©2017 Salerni et al.]

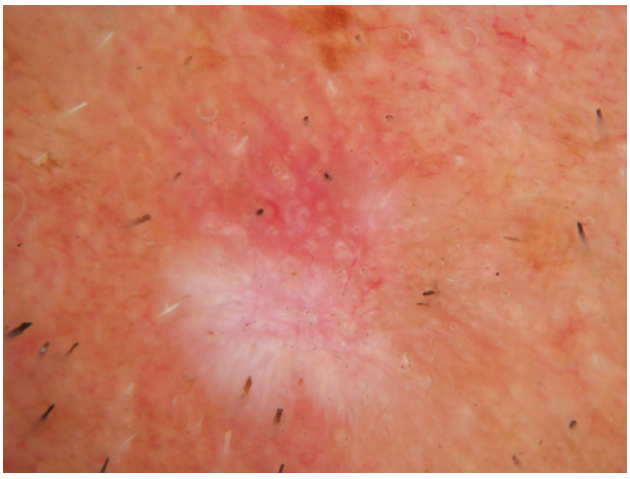

Figure 2. Dermoscopy image. A white-red structureless area with crystalline structures (short white streaks and rosettes) was observed. Focal superficial linear telangiectasias were also noted. [Copyright: ©2017 Salerni et al.]

or other procedure was reported. There were no regional lymphadenopathies.

Dermoscopy showed a shiny white-red structureless area with crystalline structures such as short white streaks and rosettes, the latter term used to designate four closely aggregated white, small dots in correspondence to a follicular opening arranged in a rhombus [5]. Focal superficial linear telangiectasias were also noted. No specific criteria for melanocytic lesion were observed (Figure 2).

Excisional biopsy was performed. Histopathology reported acanthosis, hyperkeratosis and marked cellular vacuolization. The dermis was totally invaded by a neoformation formed by a large number of intensely pleomorphic, irregular cells, some giant, with large and hyperchromatic nuclei and high mitotic index, which adopted a storiform arrangement, intermixed with thick collagen bundles (Figure 3A and B). In the immunohistochemical study, the cytokeratins and the HMB45 were negative, whereas the CD68 was marked strongly positive (Figure 3C, D and E).

Discussion

Malignant fibrohistiocytic tumors include various soft tissue sarcomas that show a spectrum of peculiar clinical-patho-

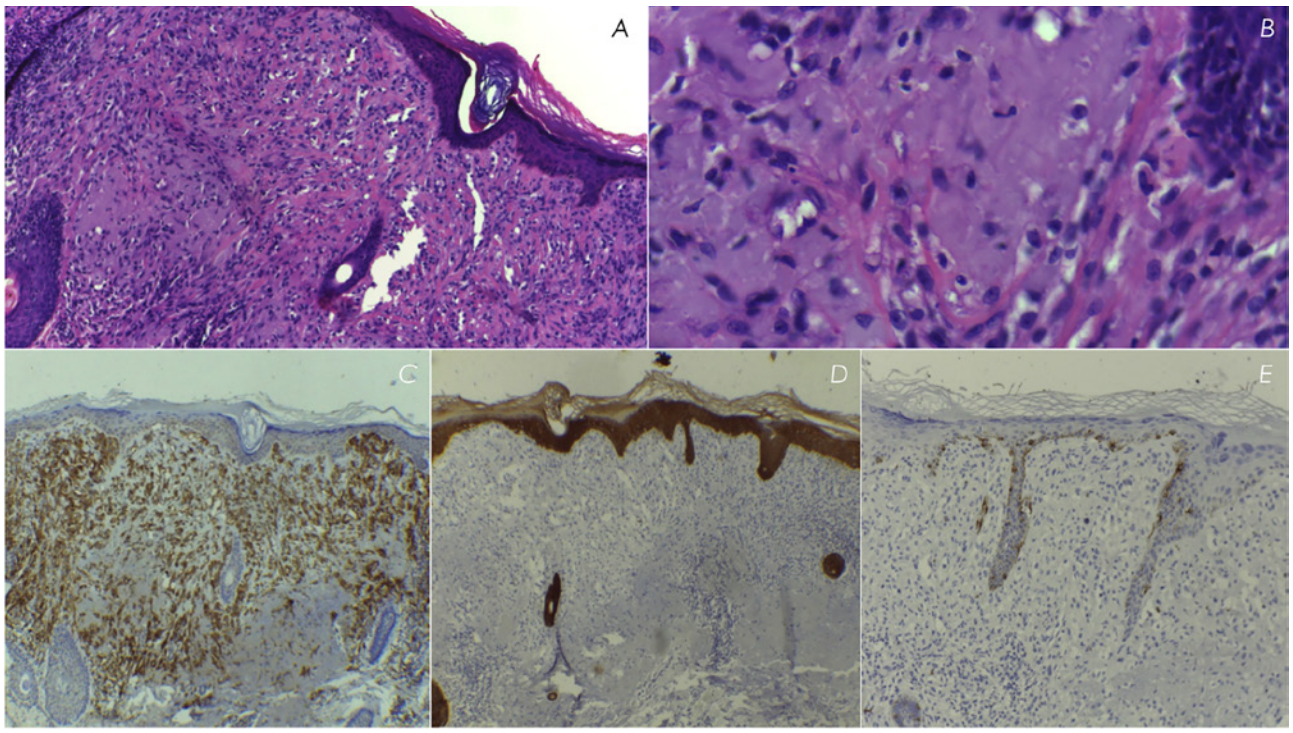

Figure 3. Non-ulcerated, mitotically active dermal fibrohistiocytic proliferation with infiltrative pattern; CD 68 positive, cytokeratin and HMB45 negative. [Copyright: ©2017 Salerni et al.]

logical findings with distinctive biological behavior. These tumors are grouped into three subtypes: atypical fibroxanthoma, dermatofibrosarcoma protuberans and malignant fibrohistiocytoma. Malignant fibrous histiocytoma (MFH) is a high-grade sarcoma that was first described by O'Brien and Stout in 1964 [6] and is currently classified as undifferentiated pleomorphic sarcoma. The term MFH is used in the medical literature to describe a high-grade pleomorphic sarcoma that affects adults. It is the sarcoma most frequently present in this age group [1].

MFH originates from pluripotent mesenchymal cells with the ability to differentiate into histiocytes, fibroblasts and myofibroblasts of the muscle or muscle fascia. The etiology of these tumors is unknown, although, as in all sarcomas, previous radiotherapy in the area may induce its occurrence. There are five histological subtypes: storiform-pleomorphic, myxoid, giant cell, inflammatory and angiomatoid. The pleomorphic storiform pattern is the most common, found in two-thirds of cases. In this variety, a large pleomorphic cell population with high nuclear atypia and mitosis is observed together with cells with fusiform morphology that adopt a "spoke wheel" or storiform pattern [7].

MFH usually affects the proximal limbs, although it has been described in the cephalic location. It can present as a cutaneous lesion in the form of a primary tumor or as metastasis from MFH at other sites. The pathogenesis is undefined, although MFH has been associated with physical, chemical and viral factors.

Approximately two-thirds of the tumors are located within the skeletal muscle, with fewer than 10% confined to the subcutis [7]. It usually presents as a slow-growing tumorous lesion, though there is no clinically characteristic presentation that allows for differentiation from other sarcomas.

The definitive diagnosis of MFH relies on histological studies, while immunohistochemistry helps establish the differential diagnosis with other entities with similar morphological patterns, such as pleomorphic varieties of rhabdomyosarcoma, leiomyosarcoma, liposarcoma, dermatofibrosarcoma protuberans, melanoma and atypical fibroxanthoma.

The dermoscopic findings were not conclusive: the presence of erythema and a shiny white-red structureless area along with short white streaks and rosettes primarily suggested a squamous tumor and less likely a basal cell carcinoma. The findings were not specific for melanocytic lesion or non-melanocytic lesion, so according to the two-step algorithm, the diagnosis of melanoma cannot be ruled out and biopsy is mandatory.

Tumor size and depth have been identified as the main prognostic factors [8]. Therefore, establishing an accurate and early diagnosis is of crucial in improving the tumor prognosis.

To our knowledge, this is the first description of the dermoscopic aspect of MFH. Dermoscopy was helpful in defining clinical management allowing for the early recognition of this tumor in an unusual presentation. MFH should be included in the differential diagnosis when assessing non-pigmented lesions with dermoscopy.

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