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CASE REPORT

Desmoplastic melanoma

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Abstract

Desmoplastic melanoma is a rare variant of malignant melanoma that accounts for 1-5% of all forms of this neoplasm. It usually affects patients in the 6th and 7th decades of life. A 42-year-old female patient with progressive scalp lesion and diagnosis of desmoplastic melanoma is described, presenting good evolution with immunotherapy using Pembrolizumab, avoiding surgical treatment.

Keywords: head and neck neoplasms; melanoma; desmoplastic; scalp.

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Introduction

Desmoplastic melanoma (DM) is a rare variant of spindle cell melanoma characterized by invasive proliferation of these cells in the dermis and varying degrees of stromal collagen deposition[1]. It is more frequently observed in male individuals aged 66 years, on average, with history of chronic exposure to the sun, which may explain its predilection for photo-exposed areas, notably the head and neck region (53.2%)[1].

Due to its extremely variable and nonspecific clinical presentation, DM represents a real diagnostic challenge. It commonly presents as a nodule, papule or plaque, hypomelanotic or amelanotic, of firm consistency, affecting the dermis or even subcutaneously, similar to other fibrous lesions, which leads to diagnostic error[2].

Histologically, it presents infiltration of spindle cells with mild to marked nuclear atypia, which invade the dermis and subcutaneous cell tissue. They are arranged in variable patterns of desmoplasia, neurotropism, and neural differentiation[1,2].

Case report

A 42-year-old female patient sought care for the appearance of a single, heterogeneous, reddish lesion on the scalp that was poorly delimited and had developed for the past three months. Computed tomography (CT) of the skull showed a poorly delimited 35mm x 15mm lesion in close contact with the skullcap. Tissue samples were collected from the affected region for biopsy and the anatomopathological examination showed a predominantly intradermal neoplasm of atypical spindle cells, with prominent multifocal

dense hyaline collagenous stroma, together with a multifocal lymphoid infiltrate. The immunohistochemical panel identified the following protein staining: multifocal positivity for S100, stronger and diffuse positivity for SOX10, positivity for KI-67 in 60% of the cells, and negativity for MART-1, leading to the diagnosis of desmoplastic invasive malignant melanoma, with Breslow thickness of 17mm and Clark level V, compatible with DM. Neoplasm staging was performed by magnetic resonance imaging (MRI) of the skull and showed two high lesions in the left superior frontal region, without contiguity, with 26mm x 20mm bone tissue. After discussion with the multidisciplinary team, a choice was made for immunotherapy with programmed death ligand 1 inhibitor (anti-PD-L1) (with an option for the use of intravenous Pembrolizumab 200 mg with a 21-day cycle) instead of surgery, due to its size. The patient presented good clinical response and progressive lesion reduction throughout the treatment, leading to disease control – Figure 1. The last positron emission tomography–computed tomography (PET-CT) showed stability of dimensions and glycolytic metabolism of the two areas of skin thickening in the skullcap in the left superior parietal and left frontal parietal regions, maximum standardized uptake value (SUV) of 1.4. As there was no suggestion of activity of the underlying disease, she was followed-up together with Clinical Oncology team.

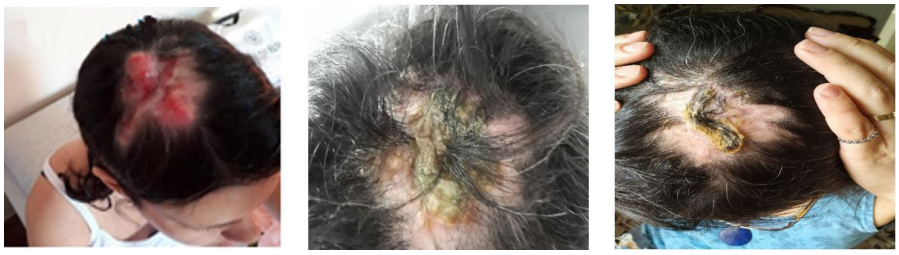

Figure 1. Evolution after 18 months of treatment (from left to right): before treatment, after 12 months of immunotherapy, and at 18 months of treatment.

Discussion

Although there is great variability in the presentation of the types of malignant melanoma, in general, its prognosis is poor, and its misdiagnosis can lead to potentially harmful handling errors associated with medical errors. Early diagnosis of malignant melanomas is, therefore, extremely important for proper treatment management and favorable survival rates[3].

Treatment is essentially surgical and consists of excision of the lesion as early as possible. Due to the more invasive behavior, particularly of the neurotropic subtypes, it is advisable to perform excision with a minimum margin of 1 cm, ideally of 2cm. Radiotherapy has been considered as an adjunct to the surgical approach aiming to reduce local recurrences[4].

Currently, immunotherapy has shown several benefits for the treatment of DM, especially in cases with metastasis, as it occurred in the case herein reported[4,5]. PD-L1 inhibitors, predominantly Pembrolizumab and Nivolumab, have been the immunotherapeutic drugs of choice due to the fact that DM

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has a high mutation load and frequent PD-L1 expression, presenting good response in evaluated studies[5].

As for prognosis, there are controversies regarding both heterogeneity of the degree of desmoplasia in the diagnosis and need for greater adaptation in the methodology of comparative studies[4,5].

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