

## Dermatomyositis Triggered by Metastatic Subungueal Melanoma: A Case Report and Review of Literature

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### Abstract

### Case Report

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized predominantly by cutaneous and muscular abnormalities that can be associated with neoplasia. The association of dermatomyositis with nail melanoma is very rare, if not exceptional. We report the case of a paraneoplastic dermatomyositis due to an undiagnosed subungueal melanoma in a 70-year-old patient.

**Keywords:** Subungueal, Dermatomyositis, muscular abnormalities.

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## INTRODUCTION

Dermatomyositis (DM) is an inflammatory disease affecting the muscles that is well recognized as a paraneoplastic syndrome. Few observations have previously reported malignant melanoma associated to DM, none of which included a subungueal melanoma.

## OBSERVATION

A 70-year-old male, without notable history, presented with pruritic erythematous and squamous lesions in the forearms, legs, neck and abdomen since 2 weeks before admission, with periorbital edema, facial telangiectasia and mouth enanthema. These dermatological signs were associated to dysphagia and myalgia with difficulties in climbing stairs, getting up from squatting position and combing his hair. The muscular weakness was bilateral, gradual in onset and progressive.

Examination of the nails found total melanonychia of the toenail of the right big toe extending over the nail folds bleeding on contact. This lesion which had progressed gradually over the past year was ignored by the patient. Dermoscopic

examination of nail bed and matrix found a multicomponent pattern made of streaks, globules, dots, structureless areas, and brown-black pigmentation along with a positive Hutchinson's sign. Laboratory findings revealed elevated muscle enzymes and LDH levels. No autoantibodies were detected. Electromyography was suggestive of myopathic process.

Pathological examination of the nail matrix biopsy found a pigmented malignant tumoral proliferation while immunohistochemistry showed cytoplasmic expression of anti HMB454 and Melan A, concluding to an ulcerated acrolentiginous melanoma (Breslow > 0.5 cm and Clark IV). Cerebral and thoraco-abdomino-pelvic computed tomography for extension work up revealed metastatic sub-tentorial brain lesions and secondary inguinal and iliac lymphadenopathy, consequently concluding to a stage IV melanoma.

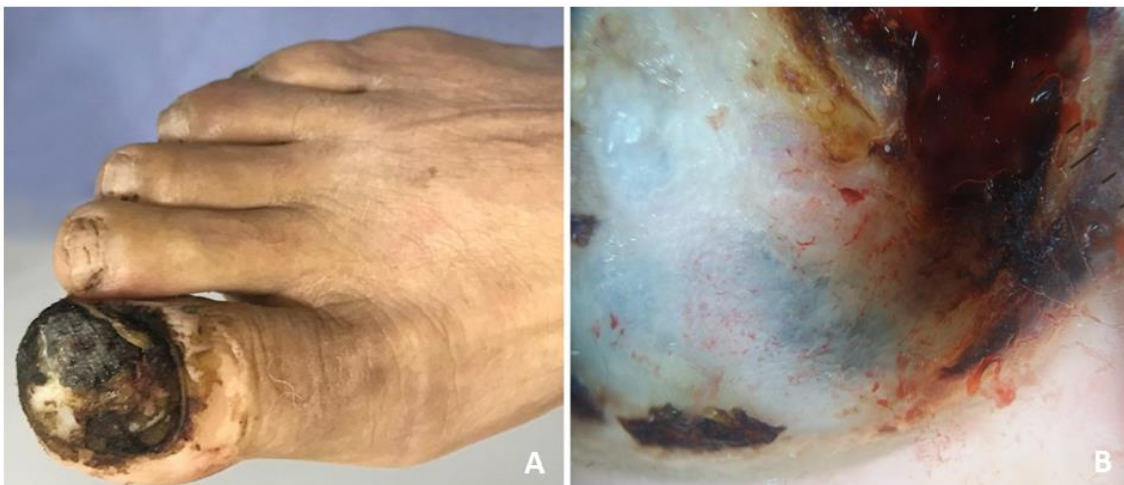
The patient was administered with prednisolone and a big toe amputation was performed. A transient improvement was noted. A recurrence occurred two weeks later resulting in fatal outcome.



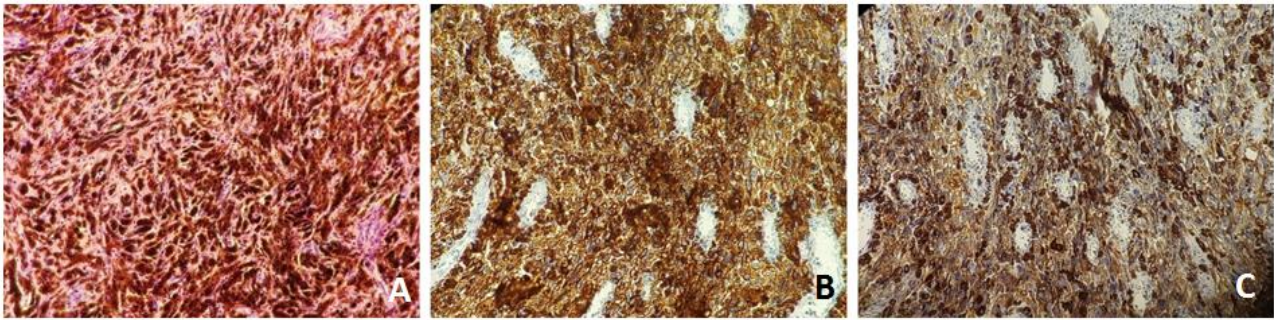
**Fig-1:** Erythematous and squamous lesions in the forearms and abdomen



**Fig-2:** Bucal mucosa enanthema



**Fig-3:** A: Total melanonychia of the big toenail extending on nail folds. B: Dermoscopic examination: Multicomponent pattern made of streaks, globules, dots, structureless areas, and brown-black pigmentation with a positive Hutchinson's sign



**Fig-4: A: Pigmented malignant tumoral proliferation (HEX20). B: cytoplasmic expression of anti HMB454. C: cytoplasmic expression of anti-Melan A**

## DISCUSSION

DM is a rare inflammatory connective tissue disorder that combines pathognomonic skin changes and muscle weakness. It is associated in 18 to 32% of cases with an underlying neoplasia appearing before, simultaneously, or after diagnosis [1]. Ovarian, gastrointestinal, lung and breast cancers as well as Hodgkin lymphomas account for 70 % of the malignancies, association to melanoma is extremely rare [1, 2]. The pathogenic mechanism remains poorly understood, but findings suggest that the relationship between myositis and malignancies is related to common expression of autoantigens [3].

The first cases of DM occurring in melanoma were published in 1960 and despite the increasing incidence of melanoma worldwide, few cases describing this association have been reported [4, 5]. No case of association to nail melanoma was found in published literature.

In a considerable literature review of paraneoplastic DM, Barnes concluded that malignancy and DM were most often discovered within 1 year of each other. He argued that this time interval permits a rational association, and the malignancy may be present in a subclinical state prior to the onset of DM [6]. According to this model, the DM process is more likely to be inducted in the presence of metastatic than primary melanoma. This can be explained by the significant tumour antigen load occurring in metastatic melanoma and consequently an increased ability for starting an autoimmune response[7].

The treatment is essentially based on non-selective immunosuppressants and in particular long-term systemic corticosteroids but especially on the etiological treatment of associated neoplasia. In some observations improvement was found in DM without immunosuppressant after cancer resection [8], but the majority of published cases of DM associated with melanoma occur at a late stage (28% of patients with stage III or IV, only 11% stage I or II), thus representing an indicator of metastatic evolution and a poor prognostic factor with a life expectancy of only a few months [5, 7, 9].

Melanoma staging has been proved to be the most important prognostic factor, regardless of the time of diagnosis. Schiller *et al.* conducted a systematic database review where they studied: the time of onset of DM in relation to diagnosis of melanoma, stage of melanoma and survival time after diagnosis of DM were recorded. They showed that the survival rates in patients with stage III melanoma are 60% at 6–12 months and 40% at 2 years and are similar to Stage III cases of melanoma in association with DM. In contrast, stage IV melanoma is associated with 1 year survival rates of 40 to 60%, while patients with DM and melanoma had a very poor prognosis and had a 1 year survival rate of 0%[4].

Part of DM cases related to paraneoplastic processes is associated with autoimmunity and oncogenesis. It was suggested that the relationship between myositis and malignancies is related to expression of auto antigens [3]. Immunosuppressive and biological treatments might also be responsible of this unusual autoimmune reaction. In the last few years, authors reported onset of DM in patients treated for melanoma with adjuvant interferon alpha and anti-PD-1 treatment or [10, 11]. The hypothesis of an incriminating both melanoma progression as well as interferon alpha therapy in the development of DM was also suggested by AI Liakou *et al.* [12].

## CONCLUSION

The prognosis is an increasingly poor in patients associating DM with neoplasia. The search for neoplasia during adult DM should be systematic. DM activity may serve as an indicator of disease progression and activity when associated with Melanoma.

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