

A rare case of late myelodysplasia cutis associated with essential thrombocythemia: A case report

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Abstract

Myelodysplasia cutis is a relatively new described entity that is characterized by cutaneous plaques and nodules representing dermal infiltration of myeloid immature non-blastic cells. It can be related to myelodysplastic syndromes or myeloproliferative disorders. It has distinct clinical and histopathological features in comparison with leukemia cutis. We report an unusual case of late myelodysplasia cutis in a male patient with essential thrombocythemia. It is only the second case reported to be related to this myeloproliferative disorder.

Keywords

Cancer, dermatology, pathology

Introduction

Myelodysplastic syndromes (MDS) are a group of hematologic malignancies characterized by clonal hematopoietic stem cells, cytopenia, and abnormal cellular maturation. In contrast, myeloproliferative disorders (MPD) are characterized by an overproduction of a cell lineage instead of a cytopenia. They share certain features with acute myeloid leukemia (AML) and chronic myeloid leukemia (CML) and can eventually progress into these entities.

The well-recognized cutaneous manifestations of MDS and MPD are diverse and include Sweet syndrome and leukocytoclastic vasculitis.^{1,2} More recently, skin lesions characterized by dermal infiltration with immature non-blastic myeloid cells have been described as a new entity called “myelodysplasia cutis.”

We report an unusual case of myelodysplasia cutis in a male patient with essential thrombocythemia.

Case report

A 65-year-old Caucasian male patient first presented to our outpatient dermatology clinic complaining of asymptomatic erythematous papules and plaques on the trunk for the previous few months. He was known only for hypothyroidism and essential thrombocythemia, the latter being diagnosed more than 10 years earlier and treated with aspirin only. At that time, a skin biopsy showed superficial and deep lymphocytic infiltrates with mucin and no atypia. A diagnosis of tumid lupus was established and the patient was initially treated

with potent topical corticosteroid cream. He was then, unfortunately, lost to follow-up.

The patient returned 3 years later. In the meantime, his family doctor had started hydroxychloroquine to try to control his skin disease, but to no avail. On physical examination, the skin lesions had changed since the initial evaluation. In fact, the patient now had more infiltrative erythematous to violaceous plaques and nodules on the upper chest and posterior trunk (Figure 1). With this evolving presentation, a skin biopsy was repeated and a computed tomography (CT) scan of thorax, abdomen, and pelvis was ordered. Histopathological analysis showed an atypical myelomonocytic proliferation CD3+, CD4+, CD68+, CD56+/-, MPO+ (myeloperoxidase), CD34-, and CD117- (Figure 2). Moreover, enlarged axillary lymph nodes and splenomegaly were noted on CT scan. A complete blood count showed thrombocytosis ($882 \times 10^9/L$). The evaluation was then completed with a myelogram, which was consistent with essential thrombocythemia, plus a bone marrow biopsy that showed myelofibrosis with a normal blast count. The

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Figure 1. Posterior trunk lesions at the time of myelodysplasia cutis diagnosis.

cutaneous diagnosis of myelodysplasia cutis secondary to essential thrombocythemia was established.

The patient was then evaluated by a hematologist-oncologist and is currently being treated with hydroxyurea. After only 6 weeks of treatment, the cutaneous lesions already improved in that they were less infiltrative (Figure 3).

Discussion

Myelodysplasia cutis, in contrast to leukemia cutis, is a relatively new described entity. It is characterized by dermal infiltration of immature non-blastic myeloid cells. A first article comparing the two entities was published about 5 years ago by a dermatology team in Paris.³ They determined the discriminating features of the two entities. Clinically, they described that patients with myelodysplasia cutis present more commonly with plaques, while those with leukemia cutis present more commonly with nodules. Our patient presented with both lesions during the course of his skin disease. Histologically, discriminating immunostaining for this diagnosis was determined by the presence of CD3+ lymphocytes and the absence of CD34, CD56, and CD117. Our patient met the criteria consistent with myelodysplasia cutis on the second biopsy, except for discrete positivity of CD56. Our patient is only the second case associated with essential thrombocythemia that has been described in the literature.⁴ During the past year, a first case of genetically proven myelodysplasia cutis was reported showing the same mutation in myeloid cells of skin and bone marrow biopsies.⁵ This suggests that the previously used non-specific term “histiocytoid Sweet syndrome” should be reserved for cases of cutaneous myeloid cell infiltration that are not associated with MDS or MPD, while “myelodysplasia cutis” would be the correct term if they are associated with these

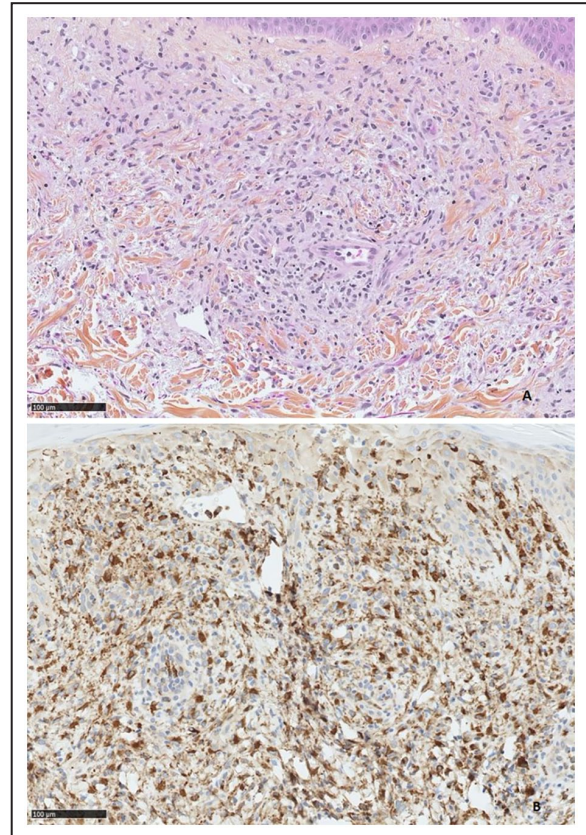


Figure 2. Histopathologic findings in cutaneous biopsy. (a) Hematoxylin–eosin staining. (b) CD68 immunostaining.



Figure 3. Posterior trunk lesions after 6 weeks of hydroxyurea treatment.

malignancies. There do not appear to be any clinical differences between the two entities but rather more a terminological distinction with regard to the etiology.

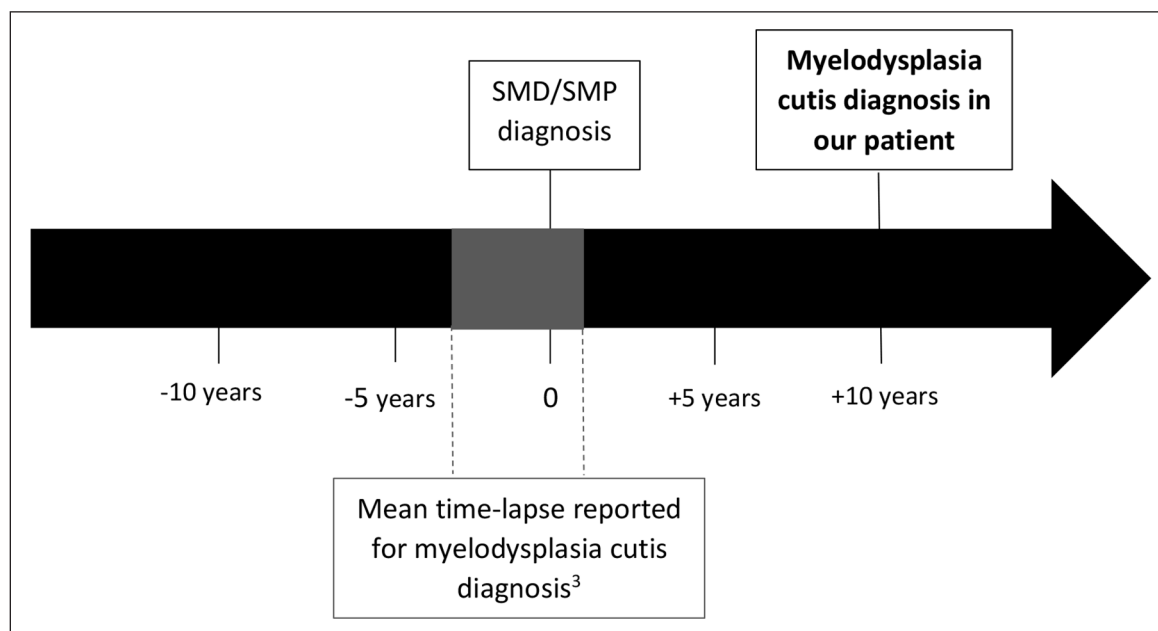


Figure 4. Timeline of hematologic and cutaneous diagnoses.

Chronologically, most reported cases of myelodysplasia cutis have occurred before the bone marrow diagnosis of MDS or MPD. However, the reverse order has also been reported for the cutaneous disease beginning at a mean of 15 months after the diagnosis of MDS (Figure 4).³ In this case, our patient's skin lesions appeared more than 10 years after the diagnosis of essential thrombocythemia, which is a longer time-lapse than the ones reported so far in the literature. Moreover, unlike most of the previously published cases of myelodysplasia cutis, our patient did not have any systemic symptoms, such as fever or arthralgia. In contrast to leukemia cutis, this new entity does not seem to affect the prognosis of the related myeloid malignancy. In the literature, patients with myelodysplasia cutis had a similar life expectancy to those with the same MDS or MPD without cutaneous involvement.³ However, the course of our patient's skin disease seemed to be consistent with the progression of his bone marrow disease, which suggests that the skin disease could be a marker of internal disease activity. Treatment of this entity is based on the treatment of the underlying bone marrow malignancy, as in our patient, who showed rapid improvement in his skin disease with hydroxyurea. Symptomatic treatment with oral corticosteroids has also been reported as being effective in some cases.⁵

In short, myelodysplasia cutis is a diagnosis to consider in every patient known to have MDS or MPD who presents with new skin lesions and could even precede the bone marrow malignancy. Our case showed that this skin disease can begin many years after the diagnosis of MPD and should not be ruled out based on a long time-lapse.

Declaration of conflicting interests

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Patient consent

Patient consent was obtained for publication of this case report.

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