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# Cutaneous Manifestation of Acute Myeloid Leukemia in an Infant

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## Introduction

Aleukemic cutaneous leukemia's are a rare form in which cutaneous lesions with myeloblasts are isolated several months before the onset of marrow involvement. These skin lesions are not uncommon in Acute Myeloid Leukemia (AML) but are exceptionally revealing. They are most often considered to have a poor prognosis. We report a case of cutaneous localization revealing acute myeloid leukemia in an 11-month-old infant.

## Observation

We report the case of an 11-month-old infant, male, with no notable pathological history, consulting the pediatric emergency department for the appearance of a right supraorbital skin nodule of 2 cm with multiple maculopapular skin lesions of purplish color located on the back, trunk, face, gums and sparing the limbs. These lesions are disposed on a non-erythematous background and are not pruriginous, all evolving for 2 weeks (Figure 1). A skin biopsy was performed showing a dermis infiltrated by a tumor proliferation arranged in diffuse sheets (Figure 2). The epidermis is separated from the infiltrate by a grenz zone that insinuates between the collagen bundles (Figure 3). The tumor cells are small to medium in size and have a lymphocytic appearance. Their nuclei are blast-like with fine chromatin and one or two nucleoli and their cytoplasm is clear or eosinophilic. Numerous figures of mitosis are observed (Figure 4, 5). Immunohistochemical complement was performed. The tumor cells express CD68, CD56, LCA, CD4 and CD15 (Figure 6). However, they did not express CD3, CD20, TDT, CD1a, chromogranin and synaptophysin. Investigation of blood involvement revealed the absence of blasts in the peripheral blood and in flow cytometry. The diagnosis of a cutaneous localization of acute myeloid leukemia (Aleukemic) was retained. The child was treated with the AML-MA 2011 protocol. The treatment by induction and consolidation allowed a good evolution and regression of the skin lesions.

## Discussion

Myeloid hemopathies are clonal proliferations of hematopoietic stem cells that can give specific skin lesions. They concern acute myeloproliferative syndromes; this means acute leukemia's de novo or secondary to the acuitization of chronic myeloproliferative syndrome (Figure 7). These lesions may occur several weeks or months before blood and marrow involvement [1].

This is especially characteristic of leukemia's with a monocytic component. In this type of disease, the histopathological examination directs the clinician towards bone marrow exploration. The lesions may present as nodules, infiltrated plaques and papules, erythematous or brownish, sometimes purpuric. Mucosal involvement, especially gingival, is classic and sometimes isolated [2].

Histologically, when skin lesions precede blood involvement, the infiltration is often

**Figure 1:** Multiple cutaneous violaceous maculo-papular lesions of the trunk and face, sparing the limbs in an 11-month-old infant.

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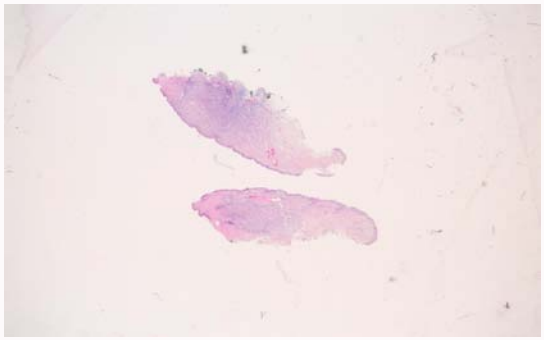

**Figure 2:** Microphotography of a skin biopsy showing a dermal infiltrate.

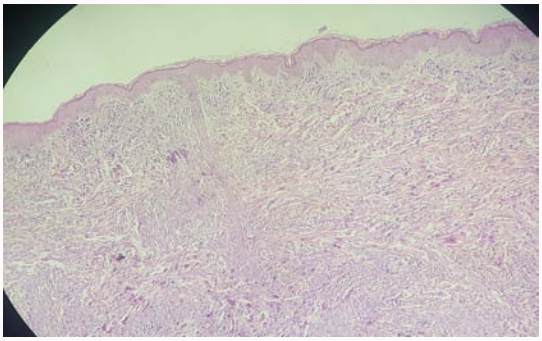

**Figure 3:** Lymphomatous proliferation in the dermis separated from the epidermis by a "grenz zone". (HE, x100).

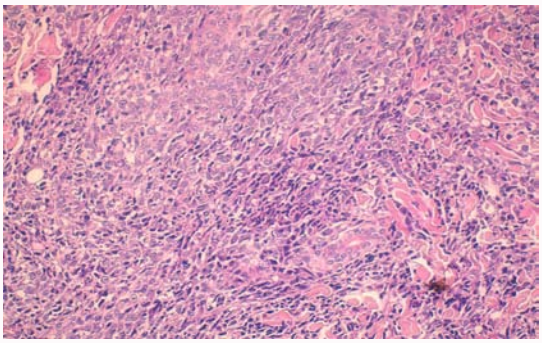

**Figure 4:** The tumor cells are small to medium sized with a blastic appearance dissociating the collagen fibers in the dermis and surrounding the adnexal structures. (HE, x200).

dermohypodermal interstitial perivascular and periannexal dissecting collagen fibers. The epidermis is separated from the infiltrate by a grenz zone. The tumor cells have a 'blastic' appearance with round or oval nuclei with fine chromatin and small visible nucleoli. Their cytoplasm is abundant in monocytic leukemia and reduced in granulocytic leukemia. Mitosis figures are frequently observed [3].

Immunohistochemically, tumor cells express markers specific to the myelocytic lineage: MPO, CD13, CD33 and/or monocytic: CD4, CD15, CD68. CD56 is expressed in about 20% of monocytic AMLs. B and T markers are negative. The Ki67 proliferation index is usually around 90% [4].

However, most often the classification of leukemia is made by biological analysis [3].

The most important histologic differential diagnosis may be with

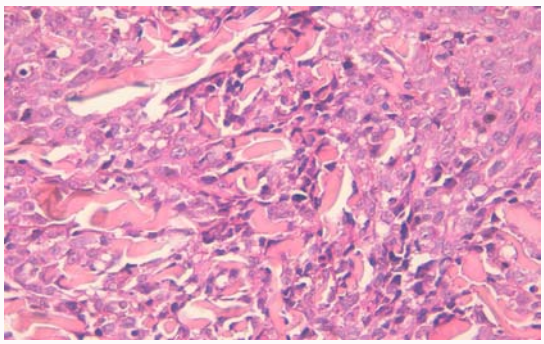

**Figure 5:** Tumor cells have blast-like nuclei, fine chromatin with one or two nucleoli and clear or eosinophilic cytoplasm. Numerous figures of mitosis are observed. (HE, x400).

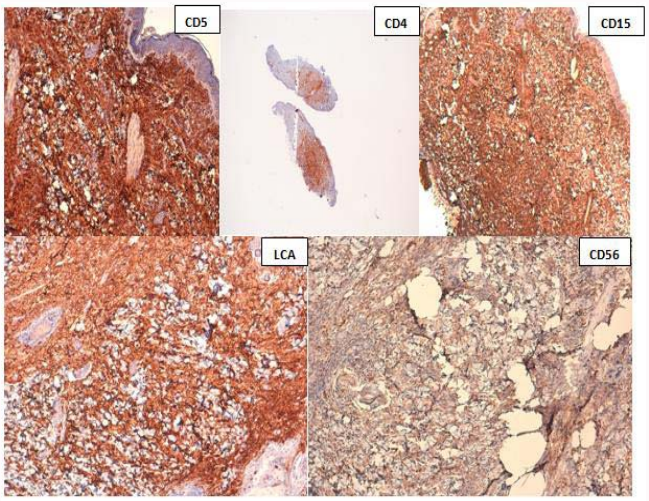


**Figure 6:** Tumor cells express CD56, CD4, CD15, ACL and CD56.

the blastic plasmacytoid dendritic cell tumor formerly known as CD4+/CD56+ hematoderma, which may be very similar in clinical presentation and morphologic appearance. Diagnosis is based on immunohistochemistry, expressing CD123, TCL1 and other plasmacytoid dendritic cell markers such as CD303 (BDCA2), MX-A and BCL11A [5].

This entity may also be confused with mature plasmacytoid dendritic cell proliferation. It is a relatively rare condition, often associated with myeloproliferative and myelodysplastic syndromes. The proliferation is made of non-blastic cells, corresponding to mature plasmacytoid dendritic cells morphologically resembling small histiocytes. It is phenotypically distinguished from blast forms by the absence of CD56 expression and the expression of granzyme B [6].

Other differential diagnoses are: B, T NK lymphomas easily eliminated by immunohistochemical study, melanoma for which PS100 positivity requires immunohistochemical complement with HMB45 and anti-Melan A/MART-1, neuroendocrine carcinomas and Sweet-histiocytic syndrome. This last one is manifested by an infiltrate of immature myeloid cells. It is often associated with myeloproliferative disorders or myelodysplastic syndromes. FISH studies have shown the presence of common genetic abnormalities in leukemic patients with Sweet-histiocytic syndrome [7]. The t (8;21) translocation is most frequently associated with acute myeloid

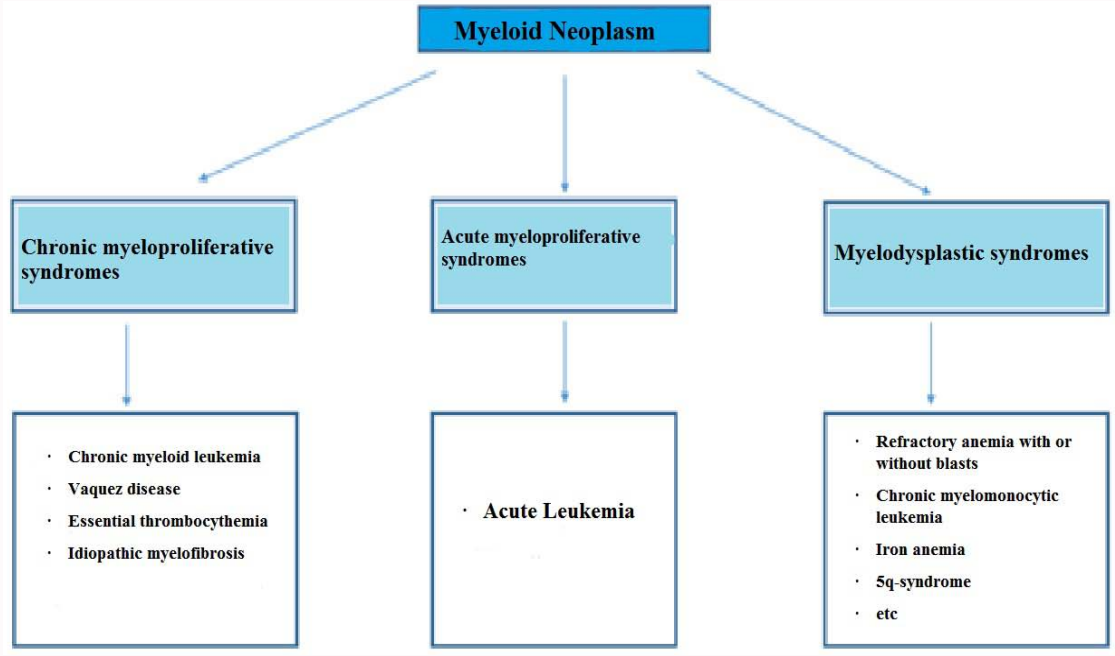


**Figure 7:** Algorithm showing the different syndromic presentations of hematological malignancies.

leukemia with skin involvement [8].

Acute myeloid leukemia with skin involvement has a worse prognosis in both adults and children. Chemotherapy is recommended if skin involvement is indicative of acute myeloid leukemia [9].

## Conclusion

This observation reminds us of the existence of cutaneous localizations in myeloid hemopathies of children that can exceptionally reveal the disease. In front of papular or papulo-nodular violaceous lesions of acute onset, histopathological examination allows the diagnosis to be made. Its evolution is very pejorative. In our case, the treatment permitted the control of the cutaneous lesions.

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