Myeloid Neoplasms

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KEYWORDS

• Myeloid • Neoplasms • Skin • Leukemia • Cutis • Acute • Chronic
• Myelomonocytic

KEY POINTS

- Myeloid neoplasms may involve the skin and may be the presenting sign of underlying bone marrow disease.
- Dermal infiltration by neoplastic myeloid cells may occur in otherwise normal skin or in sites of cutaneous inflammation.
- Leukemia cutis may precede evidence of blood and/or bone marrow involvement (aleukemic leukemia cutis).
- The classification of myeloid neoplasms has undergone major changes and currently relies heavily on genetic abnormalities.

INTRODUCTION

Myeloid neoplasms comprise a complex and heterogeneous group of hematopoietic diseases with variable prognosis.[1] The classification of myeloid leukemias has undergone major changes and currently relies heavily on genetic abnormalities (**Box 1**).[2] Cutaneous manifestations may be the presenting sign of underlying bone marrow disease.[3] A variety of patterns may occur in the skin and include cutaneous involvement by acute myeloid leukemia (AML), inflammatory dermatoses with variable association with myelodysplastic and myeloproliferative disorders, cutaneous involvement by chronic myelomonocytic leukemia, and cutaneous involvement by blastic plasmacytoid dendritic cell neoplasm.

Cutaneous Manifestations of Acute Myeloid Leukemia (AML)

Certain inflammatory dermatoses may be associated with underlying myelodysplastic/myeloproliferative disorders and include Sweet syndrome, pyoderma gangrenosum, neutrophilic eccrine hidradenitis, vasculitis, and erythema nodosum.[4] In addition, the skin may be infiltrated by leukemic cells. Myeloid leukemia cutis occasionally precedes

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Box 1
Subtypes of acute myeloid leukemia.[a]

Acute myeloid leukemia (AML) with recurrent genetic abnormalities
- AML with t(8;21) (q22;q22.1);RUNX1-RUNX1T1
- AML with inv(16) (p13.1q22) or t(16;16) (p13.1;q22);CBFB-MYH11
- Acute promyelocytic leukemia with PML-RARA
- AML with t(9;11) (p21.3;q23.3);MLLT3-KMT2A
- AML with t(6;9) (p23;q34.1);DEK-NUP214
- AML with inv(3) (q21.3q26.2) or t(3;3) (q21.3;q26.2); GATA2, MECOM
- AML (megakaryoblastic) with t(1;22) (p13.3;q13.3);RBM15-MKL1
- Provisional entity: AML with BCR-ABL1
- AML with mutated NPM1(see **Figs. 1** and **2**)
- AML with biallelic mutations of CEBPA
- Provisional entity: AML with mutated RUNX1 (see **Fig. 5**)

AML with myelodysplasia-related changes (see **Fig. 3**)

Therapy-related myeloid neoplasms

AML, NOS (not otherwise specified)
- AML with minimal differentiation
- AML without maturation
- AML with maturation
- Acute myelomonocytic leukemia
- Acute monoblastic/monocytic leukemia
- Pure erythroid leukemia
- Acute megakaryoblastic leukemia
- Acute basophilic leukemia
- Acute panmyelosis with myelofibrosis

Myeloid sarcoma

[a]Three examples are shown in Figs. 1–3, 5
Adapted from Arber DA, Orazi A, Hasserjian R, et al. The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia. Blood 2016;127(20):2392; with permission.

evidence of peripheral blood and/or bone marrow involvement (aleukemic leukemia cutis).[5]

Specific AML subtypes have been refined by focusing on significant cytogenetic and molecular genetic subgroups with variable impact on prognosis and treatment.[1] A large number of recurring, balanced cytogenetic abnormalities are recognized as subtype defining by the World Health Organization (WHO) classification (see **Box 1**).[2] Tissue infiltrates of AML (myeloid sarcoma) may represent a unique clinical presentation of any subtype of AML.[1] Myeloid sarcoma may present de novo, may accompany blood and marrow involvement, may present as relapse of prior AML, or may present as progression of a prior myelodysplastic syndrome, myeloproliferative neoplasm, or myelodysplastic/myeloproliferative neoplasm.[2] In a large series of myeloid leukemia cutis, skin lesions were de novo in 7.5%, concurrent in 26.6%, and subsequent in 60.7%.[6] Although any subtype of AML may involve the skin secondarily, acute myelomonocytic leukemia and acute monocytic leukemia are the most common.[6]

Most patients with myeloid leukemia cutis are adults, but children and neonates may also be affected, particularly in cases of acute leukemia with MLL translocations.[1] Skin lesions may be reddish-brown or violaceous papules, plaques, or tumors. Oral mucosa (especially gingiva) may be involved.[5]