

## Myeloid sarcoma

Liron Pantanowitz, MD;  
Lester D.R. Thompson, MD

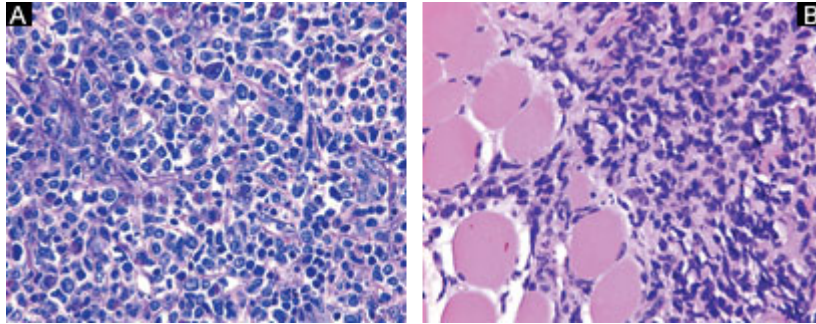


Figure. **A:** A diffuse infiltrate of myeloid leukemia cells containing fine nuclear chromatin and distinct nucleoli. Several cells contain a moderate amount of granular eosinophilic cytoplasm. **B:** Monoblasts are cells with immature nuclei and minimal cytoplasm. They are seen infiltrating into skeletal muscle.

Myeloid sarcoma (MS) is an extramedullary myeloid tumor (granulocytic sarcoma) that can occur in one of three clinical settings: (1) in patients who have a history of acute myeloid leukemia (AML), during active disease or a recurrence; (2) in patients with chronic myeloproliferative disorder or myelodysplastic syndromes, who are at increased risk of blast transformation or acute leukemia; or (3) in patients with no history of hematologic disease, although it commonly predates the development of leukemia, often within 1 year.

MS is a localized mass of myeloblasts with or without additional immature myeloid cells. *Chloroma* (*chlor* = green, *oma* = tumor) is an old clinical term that refers to the green hue identified in fresh specimens as a result of myeloperoxidase production. This green hue is lost with formalin fixation. In the head and neck region, MS has a propensity to involve the subperiosteal bones of the skull, paranasal sinuses, and orbit, as well as the soft tissues of the nasopharynx, lymph nodes, and skin. It occurs most often in pediatric and young adult patients.

MS is made up of a diffuse, fairly monotonous infiltrate of medium-size or large cells with three levels of differentiation: *blastic*, *immature*, and *differentiated* (figure):

- The blastic type is made up of myeloblasts with round-to-oval nuclei, 2 to 4 small nucleoli, fine nuclear chromatin, and a rim of basophilic, finely granular cytoplasm (sometimes containing Auer rods).
- The immature type is intermediate, made up of both blast-like cells and promyelocytes with variably lobated nuclei.
- The differentiated stage is primarily made up of promyelocytes with eosinophil and neutrophil maturation. The presence of immature eosinophils and neutrophils usually indicates the true nature of this lesion (as opposed to a malignant lymphoma).

Histochemical staining for chloroacetate esterase (Leder stain), which stains neutrophils and precursors, is usually positive, although sometimes only focally. Giemsa-stained touch preparations demonstrate cytoplasmic granules. Immunohistochemical stains (myeloperoxidase, lysozyme, CD34, nonspecific esterase, and CD68), as well as flow cytometry, are frequently required to make a definitive diagnosis.

MS is frequently misdiagnosed, especially when it precedes a diagnosis of AML by several months or even years. The differential diagnosis includes a variety of lymphomas, neuroblastoma, rhabdomyosarcoma, and Ewing sarcoma or primitive neuroectodermal tumors. When correctly diagnosed, treatment is identical to that for AML, with systemic chemotherapy and possible stem cell transplantation; local radiation is employed in the absence of bone marrow involvement.

### **Suggested reading**

Brunning RD, Matutes E, Flandrin G, et al. Acute myeloid leukaemia not otherwise categorised. In: Jaffe ES, Harris NL, Stein H, Vardiman JW, eds. World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Haematopoietic and Lymphoid Tissues. Lyon: IARC Press, 2001.

Cankaya H, Ugras S, Dilek I. Head and neck granulocytic sarcoma with acute myeloid leukemia: Three rare cases. Ear Nose Throat J 2001;80:224-6, 228-9.