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# Lytic lesion of the knee in a 27-year-old man

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A 27-year-old man complained of right knee pain and lateral knee fullness for 3 months without a history of trauma, and chronic right ankle pain for 7 months. Although he reported mild fatigue, he denied weight loss, fever, anorexia, or other constitutional symptoms. Physical examination was positive only for tenderness of the popliteal space of the right knee. Laboratory studies including complete blood count and differential were within normal limits. Bone marrow aspirate yielded slight hypocellularity.

Plain radiography of the right knee demonstrated a lytic lesion of a permeative pattern of the distal femur (*Figure 1*). Whole-body radionuclide bone scan demonstrated increased activity of the right distal femur and distal tibia and of the left lateral femoral condyle (*Figure 2*). Plain radiography of the right ankle and left knee was negative. Computed tomography (CT) of the knees revealed cortical disruption of the right distal femur with a surrounding soft tissue mass (*Figure 3*). Spin-echo magnetic resonance imaging (MRI) of the distal femurs demonstrated abnormal signal involving the bone marrow of the right distal femur extending into the surrounding soft tissues (*Figure 4*) and a similar lesion confined to a portion of the left lateral femoral condyle.

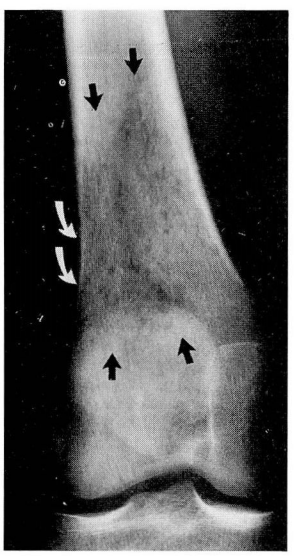

**FIGURE 1. Plain film AP radiograph of the right knee demonstrates a lesion consisting of multiple tiny lucencies of the distal femur with poorly defined margins (straight arrows) and with endosteal involvement (curved arrows).**

An open biopsy of the right distal femur and the extraosseous mass was performed. Multiple pieces of red, yellow, and tan soft tissue aggregating to 1.0 × 1.0 × 2.5 $cm^3$ were received for microscopic examination. The extraosseous mass was composed primarily of atypical immature mononuclear cells with large nuclei (*Figure 5*). Many nuclei were oval; others were angular and had indentations. Nucleoli were prominent in many of the cells. Admixed with the immature cells were

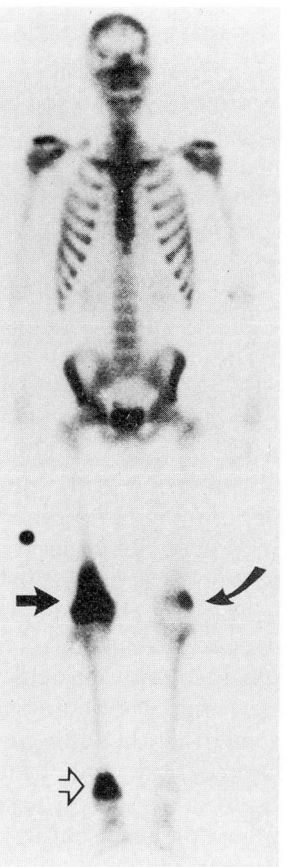

**FIGURE 2. Technetium 99m radionuclide bone scan (delayed frontal whole-body image) demonstrates increased uptake involving the right distal femur (solid arrow), right distal tibia (open arrow), and left lateral femoral condyle (curved arrow).**

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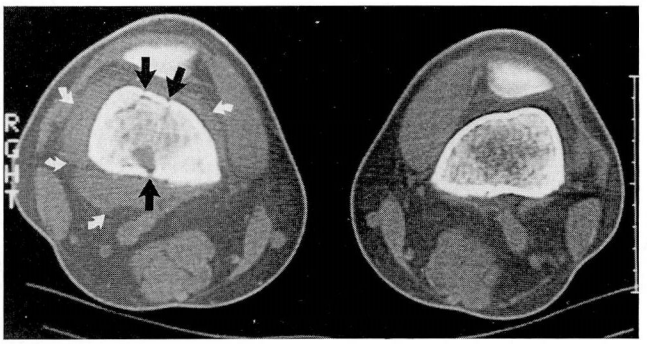


FIGURE 3. Non-enhanced CT scan of the knees reveals a lytic lesion of the right distal femur with cortical disruption (black arrows) surrounded by a soft tissue mass (white arrows).

scattered eosinophils in various stages of maturity. To confirm the suspected myeloid origin of the cells, immunohistochemical stains were used; the myeloperoxidase, muramidase, and neutrophil elastase stains were positive (*Figure* 6); Leu-4 (CD3, a pan-T-cell marker) and HLA-DR (B-cells and activated T-cells) were negative.

## DIAGNOSIS: MULTIFOCAL GRANULOCYTIC SARCOMA OF BONE

The patient underwent radiation therapy of 3,000 rads to the right distal femur, right distal tibia, and left distal femur. He developed an additional biopsy-proven lesion of the posterior chest wall during completion of radiation therapy. Due to the progression of the disease, he underwent a chemotherapy regimen of high-dose cytarabine and daunorubicin. Bone marrow aspirate again revealed hypocellularity with no evidence of leukemia. His peripheral blood studies also remained without evidence of leukemia. The patient then underwent autologous bone marrow transplantation 5 months after his diagnosis. Unfortunately, he expired from complications of *Streptococcus viridans* septicemia. Autopsy findings showed that granulocytic sarcoma was present in peripancreatic and mediastinal lymph nodes.

## DISCUSSION

Granulocytic sarcoma (myeloblastoma) is an extramedullary tumor of immature myeloid cells. Common sites include bone, periosteum, soft tissue, lymph nodes, and skin.[1] Although the tumor can occur in patients without preexisting hematologic disease, it more frequently arises in patients with acute myeloid

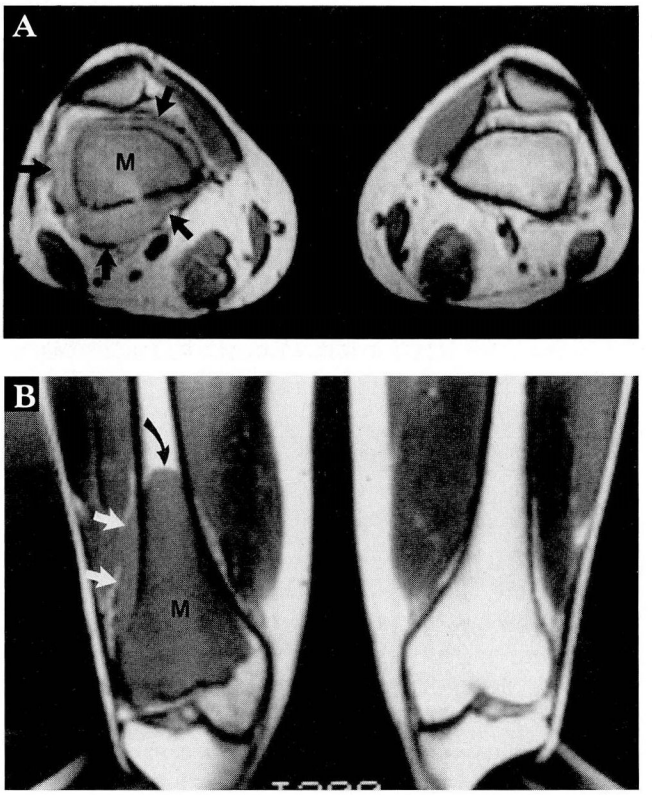


FIGURE 4. (A) Spin density intermediate weighted (TR 2000, TE 20) axial spin-echo MRI at the same level as *Figure 3*. (B) T1-weighted (TR 600, TE 20) coronal spin-echo MRI. Abnormal signal (M) replaces the normal marrow fat of the right distal femur and extends into the soft tissues (straight arrows). Note the sharp delineation of lesion extent in both, especially the proximal margin on the coronal T1-weighted image (curved arrow).

leukemia (AML) or one of the myeloproliferative disorders.

In Neiman's study[1] of 61 granulocytic sarcomas from 50 patients, 25 patients had a known myeloproliferative disorder at the time granulocytic sarcoma was diagnosed, 11 patients had known acute myeloid leukemia, and 15 patients had no known hematologic disorder. Of these 15 patients, 13 developed AML from 1 to 49 months after the original tumor diagnosis. The prognosis of patients who already have AML is not worsened by the development of a granulocytic sarcoma; in contrast, the development of granulocytic sarcoma in patients with a myeloproliferative disorder is often either a sign of a current blast crisis or the herald of AML's arrival within 4 months.[1]

The name "chloroma" has been used for these tumors because the initial color of the gross specimens

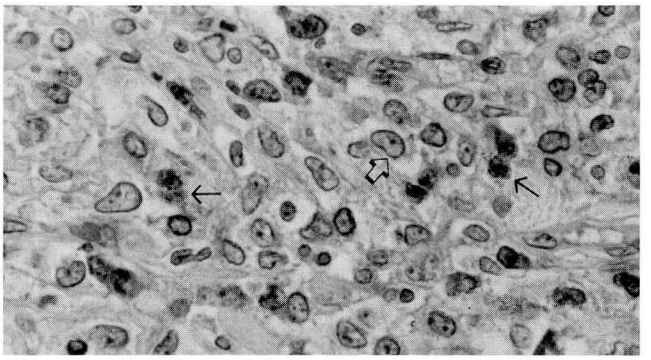

FIGURE 5. The biopsy specimen shows many immature cells with large nuclei and prominent nucleoli (single arrow). Maturing eosinophils are present (double arrows). (Hematoxylin and eosin, ×163)

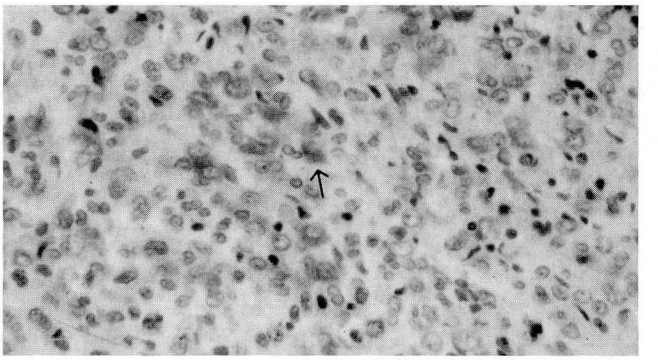

FIGURE 6. The neutrophil elastase stains the cytoplasm of neutrophils and their precursors (arrow). (Neutrophil elastase stain, immunoperoxidase technique, ×100)

is often greenish, due to the presence of verdoperoxidase, a myeloperoxidase in the cells.[1] This color fades with exposure to air. Not all tumors are green, however, and chloroma is no longer the preferred term.

There are three stages of differentiation: blastic, immature, and differentiated. The blastic tumors consist mostly of myeloblasts, with scant evidence of differentiation to promyelocytes. Eosinophils are typically absent. The immature variety contains both myeloblasts and promyelocytes; eosinophilic myelocytes are typically present. A differentiated tumor has promyelocytes and later stages of granulocytic development; eosinophilic myelocytes are plentiful.[2] The degree of differentiation has no bearing on prognosis.

These myeloid tumors can resemble tumors of lymphoid origin. Traditional stains such as myeloperoxidase and chloracetate esterase can confirm the myeloid nature of the tumor; immunoperoxidase stains such as neutrophil elastase or lysozyme (muramidase), as well as monoclonal antibodies for B cells and T cells, can exclude a lymphoid origin.[3]

The radiologic evaluation of the patient with a suspected osseous lesion should always begin with the plain radiograph. In some cases, especially those involving benign lesions, the findings may be so characteristic that additional evaluation is unnecessary. However, if additional imaging is needed, radionuclide bone scintigraphy is useful in detecting additional lesions; its advantages include the capability for whole-body imaging and an increased sensitivity compared to film radiography. (It is commonly accepted that 30% to 50% of trabecular bone must be destroyed before a lesion is evident by plain radiography, whereas a bone scan can demonstrate a loss of 5% to 15%.) CT and MRI have both been shown to be valuable in the evaluation of bone and soft-tissue tumors, with MRI being superior in delineating the extent of neoplasms and in visualizing marrow abnormalities.[4,5]

The patient's age, the anatomic site of origin of the lesion, the focal or multifocal nature of the process, and the radiologic characteristics of the lesion are all factors in the differential diagnosis. The differential diagnosis of this multifocal lytic process involving the distal long bones of a young adult included lymphoma, metastatic disease, multicentric osteomyelitis, lytic osteogenic sarcoma, round cell sarcoma, and (less likely) granulocytic sarcoma.

The radiographic appearance of granulocytic sarcoma involving osseous structures has been described as a lytic lesion which may be associated with a periosteal reaction and with an adjacent soft tissue mass. Lesions have been most commonly reported in the skull, spine, ribs, sternum, and pelvic bones.[6] These lesions are unrelated to the common radiographic manifestation of osteopenia in patients with leukemia which may or may not be associated with leukemic cell infiltration.[7] The CT and MRI findings of granulocytic sarcoma involving the appendicular skeleton have not been previously reported.

The plain film findings in this case (*Figure 1*) are similar to those previously described in granulocytic sarcoma involving bone. The bone scan (*Figure 2*), because of its increased sensitivity, demonstrated additional lesions of increased radiopharmaceutical uptake, despite normal radiographs. The unenhanced CT scan demonstrated slightly increased attenuation of the marrow space related to the replacement of normal low

attenuation fatty marrow by the tumor mass, demonstrated cortical disruption consistent with the lytic process, and demonstrated the soft tissue mass surrounding the distal femur with attenuation similar to muscle. Spin-echo MRI, which remains nonspecific despite its high level of sensitivity, demonstrated replacement of normal fatty marrow by a mass lesion extending into the surrounding soft tissues. The lesion exhibited low signal similar to muscle on T1-weighted images (*Figure 4, A*), high intermediate signal lower than fat on intermediate-weighted images (*Figure 4, B*), and mixed intermediate and high signal on T2-weighted images. The MRI findings are similar to those generally described for osseous involvement by neoplasm.[8]

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