International Journal of Surgical Pathology 11(4):271–282, 2003

Myeloid Sarcoma: Clinical and Morphologic Criteria Useful for Diagnosis

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Extramedullary accumulation of myeloblasts or immature myeloid cells form tumors called myeloid sarcoma in the WHO classification. Such tumors develop in lymphoid organs, bone (skull, orbit, etc.), skin, soft tissue, various mucosae and organs, and the CNS. They may precede or occur concurrently with acute myeloid leukemia, or reveal blastic transformation of chronic myeloproliferative disorders or myelodysplastic syndromes. They may also reveal relapses in treated patients. They are constituted by a diffuse infiltrate made up of medium-to-large cells. The cells are difficult to identify. Imprints are very useful. Immunohistochemistry can help diagnose and distinguish four variants: granulocytic myeloperoxidase (MPO+, CD 68+ [KP1±, PGM1-] lysozyme+, CD 34±), monoblastic (MPO-, CD 68+, [KP1+, PGM1+] lysozyme+, CD 34-), myelomonoblastic (MPO-, CD 68+, [KP1+, PGM1+] lysozyme+, CD 34-), or megakaryoblastic (positivity for factor VIII, CD 61, CD 31). Immunohistochemistry sometimes demonstrates expression of CD 43, CD 7, CD 79a, and CD 56 (particularly the monoblastic variant with t[8;21]). Recently the demonstration of CD 99 and CD 117, which can now be done on paraffin sections, may be useful to identify blasts of granulocytic origin. The diagnosis is missed in about 50% of cases when immunohistochemistry is not used. Patients with myeloid sarcomas should be treated in the same way as patients with acute myeloblastic leukemia. Disease progression and prognosis are similar for the two conditions. *Int J Surg Pathol 11(4):271–282, 2003*
Key words: myeloid sarcoma, granulocytic sarcoma.

The term *myeloid sarcoma* in the recent WHO classification [1] describes a lesion constituted by a tumor mass made of myeloblasts or immature myeloid cells, in an extramedullary site or in bone. This term has been proposed to replace all the other names that have been used, including *granulocytic sarcoma* [2], *extramedullary myeloid tumor*, *extramedullary myeloblastic tumor*, *myelosarcoma*, *myeloblastoma*, and *chloroma*. The term proposed by the WHO classification (2001) emphasizes that these tumors are composed of blastic, immature, or differentiated cells belonging to one of the three main hematopoietic cell lines, which arise and mature in the bone marrow.

Methods for Diagnosis

One of the most important diagnostic techniques uses imprints stained with May-Grünewald-Giemsa's solution or Wright's solution (Fig. 1). This is the easiest way to reveal the morphology of blasts, allowing elimination of the diagnosis of soft tissue tumors, such as Ewing's sarcoma. Histologic sections after formalin fixation and paraffin embedding

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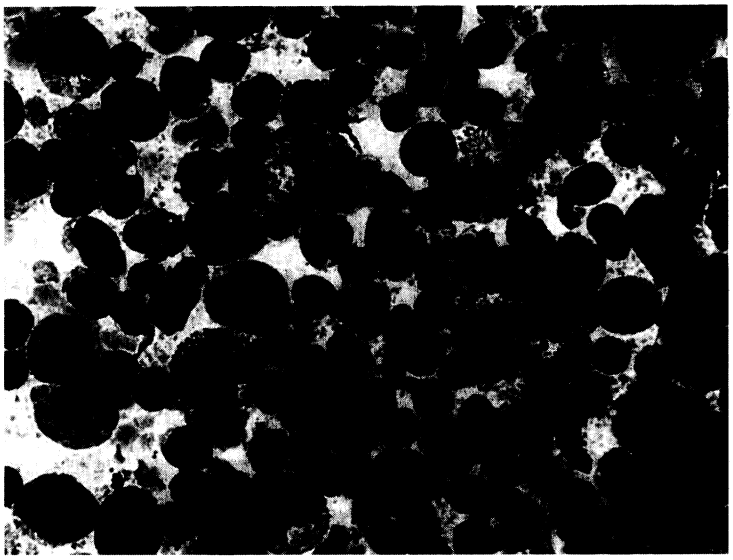

Fig. 1. Nodal myeloid sarcoma.Imprint (May-Grünewald-Giemsa). Numerous large and medium-sized cells with the morphology of blastic and immature myeloid cells dispersed between small lymphocytes.

are usually stained using hematoxylin and eosin, PAS, and, in particular, Giemsa. This facilitates recognition of blasts of hematopoietic origin.

Cytochemistry can still be useful to demonstrate myeloperoxidase (MPO) activity on smears or naphthol-ASD-chloracetate-esterase activity (with Leder stain) on paraffin-embedded tissue after fixation by formalin but not by Bouin's solution. Both stains are very useful to demonstrate the granulocytic origin of neoplastic cells.

Immunohistochemistry is essential to confirm the diagnosis of myeloid sarcoma. The following antibodies are the most useful: CD 45 (demonstrating the leukocyte origin of neoplastic cells), CD 15 (a marker for the granulocytic series), CD 68 (the accepted marker for cells from the monocytic series despite its absence of specificity). Staining for MPO and lysozyme is very informative. CD 99 and CD 117 can now be demonstrated on paraffin sections in some cases of acute myelogenous leukemia and myeloid sarcoma. CD 34 demonstrates immature blasts. Myeloid blasts may aberrantly express one or two B-cell-associated (e.g., CD 20, CD 79a), or NK/T-cell-associated (e.g., CD 2, CD 3, CD 7, CD 56, CD 57) markers. In a few patients, coexpression of myeloid markers and B or NK/T cell markers suggests biclonal proliferation. CD 43, which is today less used, is often positive. CD 56 should also be interpreted with great caution, owing to its possible expression by some small round cell sarcomas. Glycophorin C and blood group antigens are good markers for the erythroblastic series. CD 31, CD 61, and factor VIII suggest a megacaryocytic/blastic origin.

Other diseases can mimic myeloid sarcoma, and therefore other antigens have to be used to eliminate the diagnoses of undifferentiated carcinoma, melanoma, or small round cell sarcoma. Tryptase is very useful to eliminate the diagnosis of mast cell neoplasia.

Immunophenotyping of cell suspensions by flow cytometry is a very useful technique. Previously, electron microscopy was used to detect the typical granules found in granulocytes. Cytogenetic studies are also very useful and essential for patients presenting with a solid tumor in which hematopoietic cell proliferation is suspected.

Clinical Presentation

Myeloid sarcoma occurs either as a single tumor or sometimes in multiple nodular masses of various size [2–4]; in different tissues or organs such as skin, gum, lymph node, mammary glands [5]; female genital tract [6]; or testis (see Table 1). Associated symptoms depend on the site of tumor and include serous effusion [7,8], superior vena cava compression [9], bleeding due to digestive tract involvement, orbital mass with exophthalmos due to orbital localization [10,11], "spontaneous" rupture of the spleen, paraplegia due to epidural infiltration [12,13], various neurologic symptoms due to CNS

Table 1. Myeloid Sarcoma—Sites of Involvement

Lymphoid tissue: lymph node, tonsil, spleen, thymus
Subperiostal bone: skull, orbit (classical presentation), sternum, ribs, vertebrae, pelvis, etc.
Skin
Soft tissue: muscle, mediastinum, etc.
Mucosae: gastrointestinal tract, urinary tract, mouth, larynx, etc.
Different organs: breast, kidney, lung, testis, ovary, uterus (cervix), genital tract, prostate
Central nervous system and epidural infitration
Serosal cavities

involvement [14], and urinary retention due to prostatic involvement. Myeloid sarcoma can occur at any age but is most common in young adults and children and is more frequent in men than in women. Between 1985 and 2002, more than 300 case reports have been published. Four different types of clinical presentation can be distinguished as follows:

1. Myeloid sarcoma can occur in patients with known acute myeloid leukemia in the active phase of the disease. This can be observed in patients with acute myeloblastic leukemia. Tumor localization in the skin or mucosae, which can often be multiple, seems to be observed more frequently in patients with acute monoblastic or myelomonoblastic leukemia. Acute megakaryoblastic or erythroblastic leukemia can be associated with tumor formation. The diagnosis is usually easy to make and the prognosis is the same as for acute myeloid leukemia.

2. Myeloid sarcoma may develop in patients with a chronic myeloproliferative disorder (chronic idiopathic myelofibrosis, essential thrombocytemia, polycythemia vera, chronic myelogenous leukemia) or a myelodysplastic syndrome [15]. The tumor may be the first manifestation of blastic transformation. The incidence of myeloid sarcoma in patients with chronic myeloid leukemia (CML) was studied by Specchia et al [16] in 1996. Of 235 patients presenting CML, 91 showed a blastic crisis and 15 of these (16%) had an extramedullary blastic tumor. In 11 of these 15 cases (73%) the morphology was that of a myeloid sarcoma while four cases (27%) represented a lymphoblastic tumor (three with a B-cell phenotype and one with a T-cell phenotype).

3. Myeloid sarcoma may be the first manifestation of relapse in patients previously treated for primary or secondary acute leukemia, particularly after bone marrow transplantation [17,18] or in patients with chronic myelogenous leukemia following allogenic stem cell transplant [19,20].

4. Myeloid sarcoma may occur in healthy patients who have no other signs of acute myeloblastic leukemia (normal peripheral blood leukocyte count and no blasts in the bone marrow). In this situation, the diagnosis is often missed [21]. A typical form of acute myeloblastic leukemia may occur after an interval of weeks, months, or even years [21]. Although there is no peripheral blood and bone marrow involvement, patients with myeloid sarcoma should have the standard treatment for acute myeloid leukemia [21].

Macroscopy

On macroscopic examination, neoplastic tissue usually appears firm and whitish in color, with a fish-flesh appearance and consistency. In some cases, the fresh tumor tissue is a peculiar green color, owing to the presence of myeloperoxidase, fading rapidly after air exposure or formalin fixation. The term *chloroma* was given to these tumors, particularly those in the orbital bones, because of the green color observed. Large masses may contain necrotic and hemorrhagic areas.

Histopathology

The topography of the infiltrate is monotonous and always diffuse. The infiltrate can be massive, obscuring and destroying the normal tissue, or minimal with persistence of normal architecture. In the lymph node, for example, the infiltrate can be massive, destroying the architecture, or more limited (Fig. 2), infiltrating a part of the cortex, in particu-

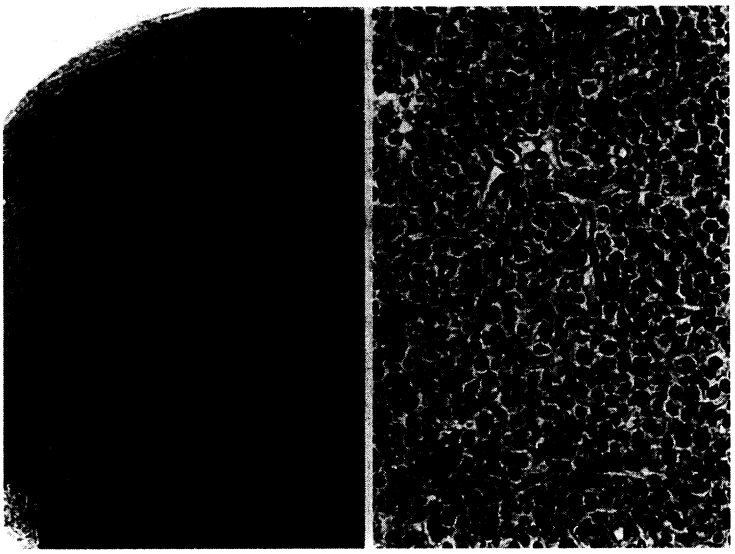

Fig. 2. Nodal myeloid sarcoma (H&E). To the left, partial infiltration between the follicles and along the sinuses (pale areas) with persistence of normal lymphoid tissue (dark areas).To the right, the myeloid cells have a clear round or oval nucleus contrasting with the dark chromatin of the nuclei of the lymphoid cells.

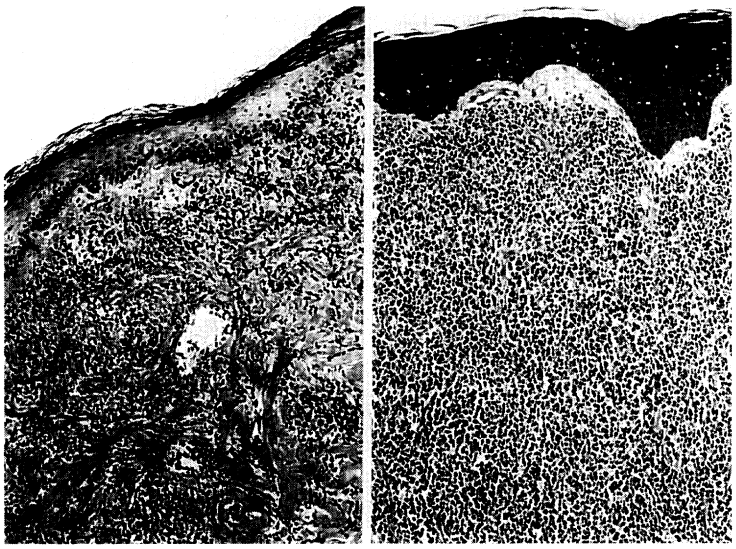

Fig. 3. Cutaneous myeloid sarcoma (H&E). To the left, the myeloid cells infiltrate the dermis between the collagen fibers and around the vessels. To the right, they realize a massive infiltrate destroying the dermis.

lar along the sinuses, between the follicles, and sometimes the sinuses. In the spleen, the red pulp is predominantly infiltrated, with persistence of the white pulp. In the skin (Fig. 3), the infiltrate can be massive, obscuring the organization of the dermis, with destruction of the adnexal glands. Skin infiltration can also be more limited (Fig. 3), occurring only in the dermis, with cells present between bundles of collagen. In all the sites involved, perivascular accumulation of tumor cells is obvious, with occasional invasion of the blood vessel walls. In some cases, areas of necrosis can be seen that sometimes can be extensive. Fibrosis may develop in some tumors.

The morphology of the neoplastic cell population is highly variable. The cells are medium-sized to large (Fig. 1), often with a monotonous pattern. Some are composed of blastic cells with a round or

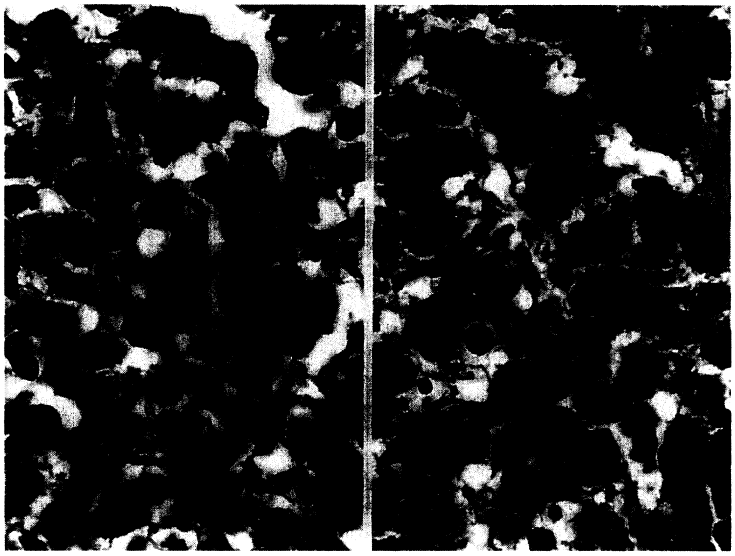

Fig. 4. To the left, cutaneous myeloid sarcoma (H&E), monoblastic variant, showing slight irregularities of the nuclei. To the right, vaginal myeloid sarcoma (H&E), myelomonoblastic variant, showing a high number of apoptotic cells.

ovoid nucleus, presenting either a large centrally situated nucleolus or medium-sized nucleoli and free or vesicular chromatin. Their cytoplasm is scant or medium-sized, and amphophilic or basophilic on Giemsa staining. Often no granules are recognizable. The mitotic count can be high. In some cases, the cells show a clear ovoid or kidney-shaped nucleus, with clear chromatin and small nucleoli. The large cytoplasm is faintly grey-blue on Giemsa staining, and there are a variable number of specific granules (mostly neutrophilic). Some of the cells may resemble promyelocytes. In rare cases of myeloid sarcoma constituted by cells belonging to the eosinophil series [22], numerous crystalline inclusions can be seen as clusters of pointed needle-like crystals either in foci of necrosis or within macrophages. They are negative for chloracetate esterase and myeloperoxidase and represent Charcot-Leyden crystals [23].

Nuclei with irregular contours (Fig. 4) with a notched or band-like pattern are often seen in monocytic cells. The association of blasts of various sizes and of larger cells with irregular multilobular nuclei often with dense chromatin, favors the diagnosis of megakaryoblastic proliferation [24]. Cases involving erythroblasts seem to be extremely rare.

Hence, in the majority of the cases, myeloid sarcomas are constituted by blastic or immature cells. In some cases, more mature myeloid cells such as promyelocytes or myelocytes can be observed. Even mature granulocytes, mostly eosinophils, may be associated. Rarely, a starry sky pattern is made up of dispersed reactive histiocytes often with phagocytosis of apoptotic cells described as "tingible bodies."

Different variants can be defined according to the predominant cell types. It has been proposed (1) to distinguish four different granulocytic variants: 1) a blastic variant with predominance of myeloblasts, (2) an immature variant constituted by a mixture of myeloblasts and promyelocytes, (3) a differentiated variant constituted by some myeloblasts associated with promyelocytes and more mature granulocytes, and (4) a variant with intracytoplasmic Auer bodies, most often occurring in acute transformation of myelodysplasia. On the other hand, monoblastic and monocytic variants can be recognized, constituted by monoblast or sometimes by cells with notched or band-like nuclei. The association of myeloblasts with more mature granulocytic cells such as myelocytes and cells with a monoblastic appearance is interpreted as a myelomonocytic variant (Fig. 4).

The association of blasts and atypical megakaryocytes (Fig. 5) or micromegakaryocytes (medium-sized cells with hyperchromatic polylobated nuclei) favors the diagnosis of acute megakaryoblastic cell proliferation [24].

Bilineage or trilineage variants are rare. They are made up of myeloblasts, myelocytes, megakaryoblasts, or erythroblast precursors. These variants are most often seen in the acute transformation of chronic myeloproliferative disorders.

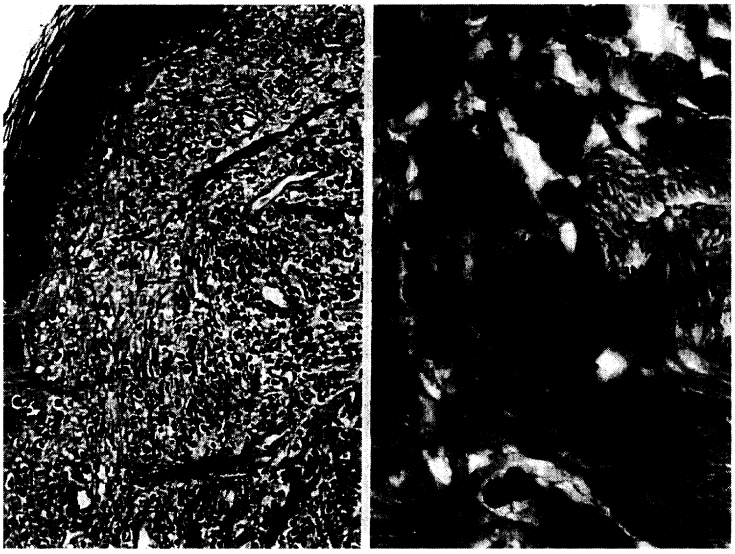

Fig. 5. Cutaneous myeloid sarcoma, megacaryoblastic variant (H&E). To the left, predominantly perivascular infiltrate in the dermis. To the right, notice the large size and the nuclear pleomorphism of the neoplastic cells.

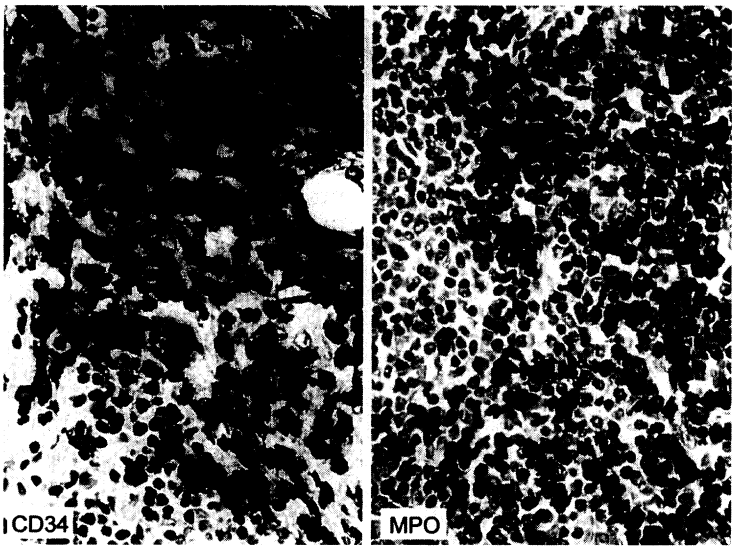

Fig. 6. Immunohistochemistry in myeloid sarcoma. To the left, in a cervical lymph node, myeloblastic variant, the neoplastic cells express CD 34. To the right, retroperitoneal mass, myelomonocytic variant, many cells are positive for MPO.

Immunohistochemistry

Immunohistochemistry is now essential to diagnose myeloid sarcoma and to try to distinguish among the different variants [25,26]. Many patients with myeloid sarcoma express only a few granulocytic or monoblastic markers, and sometimes they express some aberrant markers such as B-cell-, T-cell-, or NK-associated antigens.The expression of CD 45 demonstrates the leukocytic origin of the neoplastic cells; however often it is not expressed. MPO, a valuable marker, is positive in most myeloblastic variants (Fig. 6) as well as in some cells of myelomonocytic variants. Lysozyme is frequently expressed in monoblastic variants, but granulocytes at different phases of maturation may also be positive. CD 15 is expressed mainly by mature cells belonging to the granulocytic series and also by some histiocytes and monocytes. CD 68 is a nonspecific antigen of monocytes and macrophages recognized by different antibodies with

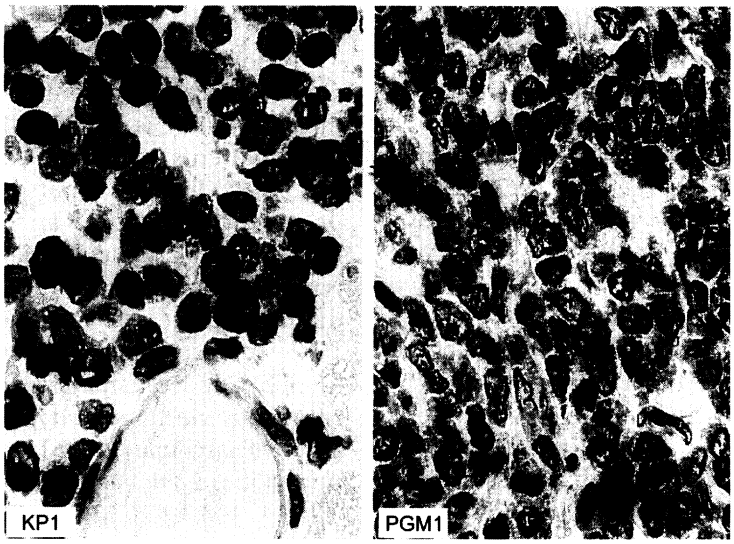

Fig. 7. Myeloid sarcoma of the intestine. Immunohistochemistry. Comparison of 2 antibodies recognizing antigens belonging to CD 68. To the left, the neoplastic cells express KP 1. To the right, the same cells are negative for PGM 1. This case, which was CD 34, MPO, and lysozyme positive, relapsed as an acute myeloblastic leukemia with maturation (FAB: M2).

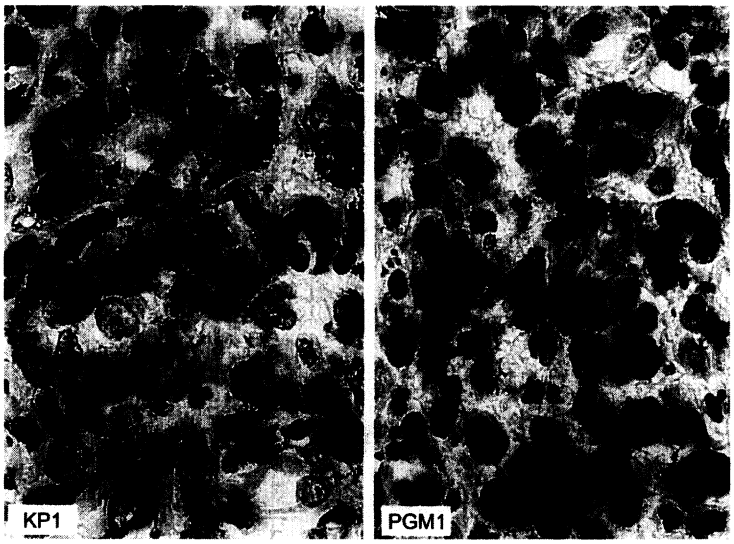

Fig. 8. Cutaneous myeloid sarcoma. Same comparison as Fig. 7. In this case, the blastic cells are positive for both KP 1 and PGM 1. This case was diagnosed as an acute myelomonocytic leukemia (FAB: M4).

different staining patterns. For example, KP 1 is expressed by cells of the monocytic and granulocytic lineages (Fig. 7), whereas PGM 1 is more specific for the monocytic series (Fig. 8). Recently two antigens expressed by cells of the granulocytic series could be demonstrated on paraffin sections. CD 117 (c-Kit) is expressed in some acute myelogenous leukemias. This antigen is also present on mast cells. CD 99 is expressed by a significant proportion of myeloid sarcoma [27], mostly in cases classified as FAB M1 or M3).

A variable percentage of blasts (Fig. 6), corresponding to immature or nondifferentiated cells, are positive for CD 34. TdT is rarely positive and only in the most blastic variants. CD 43 is often positive, and this should not be interpreted as in favor of a T-cell origin of the neoplastic cells.

Unexpected or aberrant expression of variable antigens can be observed in association with one or more granulocytic or monoblastic markers. CD 30 expression has been reported in a few cases [28]. Some blasts of myeloid sarcoma may express one or more B-cell-associated (i.e., CD 20, CD 79a) or NK/T-cell-associated antigens (i.e., CD 7, CD 2, CD 4). CD 56 is frequently expressed by monoblastic or myelomonoblastic variants [4,14,29,30], particularly in patients presenting t(8;21).

The coexpression of CD 7, CD 56, and sometimes CD 3 defines an entity called "CD 7 and CD 56 positive, myeloid/NK cell precursor acute leukemia," which often presents with localization in the skin or other extramedullary tumors. This distinct entity is now interpreted as a form of leukemia constituted by the proliferation of type 2 dendritic cells [31].

Megakaryoblastic cells (Fig. 9) are characterized by the expression of factor VIII, CD 61, and CD 31 [24]. Glycophorin C and/or blood group proteins occur in rare erythroblastic variants.

Thus immunohistochemistry on paraffin sections helps to identify different variants more precisely than use of morphology alone. For example, with use of only four antibodies, it has been proposed to distinguish some variants of myeloid sarcomas (see Table 2).

Some other markers can be studied by using frozen sections, for example, CD 13 or CD 33, for granulocytic variants and CD 14, CD 116, and CD 11c for monoblastic variants.

Genetics

Myeloid sarcomas have been observed to be associated with certain chromosomal abnormalities. The most frequent is t(8;21)(q22;q22), occurring in some patients with acute myeloblastic leukemia [4,11,32]. One case report noted the simultaneous occurrence of t(8;21) and trisomy 8 [3]. Acute myelomonoblastic leukemia can also be associated with eosinophils and can have either the gene sequence inv(16)(p13;q22) or t(16;16)(p13;q22) [33]. Acute myeloblastic leukemia in children or monoblastic leukemia associated with 11q23/MLL rearrangement is also likely to predispose to myeloid sarcoma [34–36]. Other chromosomal abnormalities have been reported [37], e.g., t(12;13), t(10;11) [38,39], t(12;22), t(3;4) [40], and the AML 1/ETO fusion gene [41].

Diagnosis

The correct diagnosis of myeloid sarcoma should be quickly made and patients should be treated rapidly with protocols similar to those used in acute myeloid leukemia. The diagnosis is relatively easy in patients with an established history of acute myeloblastic leukemia and/or myelodysplasia or chronic myeloproliferative disorders. When there is no medical history of these disorders, the diagnosis is often missed. Pathologists should consider this di-

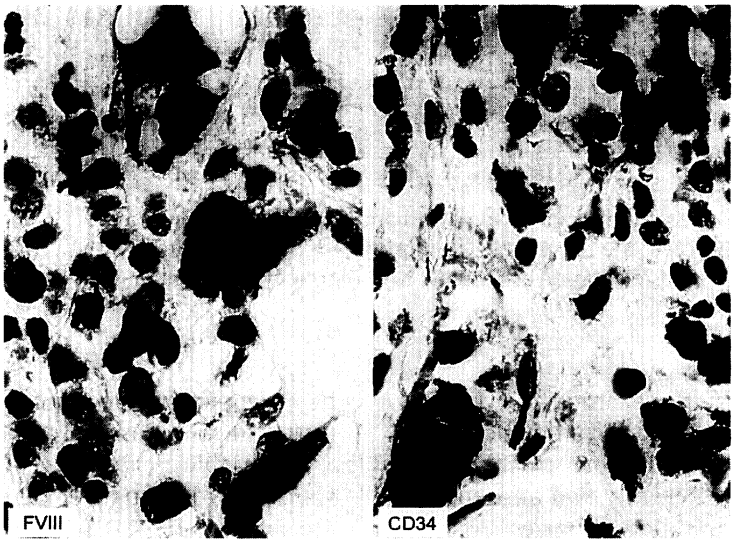

Fig. 9. Same case as Fig. 5. Immunohistochemistry. To the left, the large megacaryoblastic cells express the factor VIII. To the right, the large cells as well as some smaller ones are positive for CD 34.

Table 2. Myeloid Sarcoma: Immunophenotype of Different Variants on Paraffin-Embedded Tissue

Granulocytic variant: MPO+, CD 68+/- (KP 1+/-, PGM 1, KP 1+/-, PGM 1-) lysozyme+, CD 34+/-
Monoblastic variant: MPO-, CD 68+ (KP 1+, PGM 1+, KP 1+, PGM 1+), lysozyme+, CD 34-
Myelomonoblastic variant: MPO+/-, CD 68+ (KP 1+, PGM 1+, KP 1+, PGM 1+), lysozyme +/-, CD 34+/-
Megakaryoblastic variant: Factor VIII+, CD 31+, CD 61+
Erythroblastic variant: Glycophorin C+, blood group antigens

agnosis when there is proliferation of medium-to-large cells with round, ovoid, or irregular nuclei. The differential diagnosis is often difficult and the following diseases should be considered:

1. *Non-Hodgkin's lymphoma.* Myeloid sarcoma could be difficult to distinguish from medium-sized or large cell lymphomas. Analysis of morphology alone can be used to eliminate first some precursor cell lymphoma, then Burkitt's lymphoma, and some peripheral NK/T-cell lymphomas, mostly those constituted by a majority of medium-sized cells and associated with eosinophils. The differential diagnosis with large cell lymphoma of B-cell or T-cell type can be really difficult. Therefore, immunohistochemistry is required to distinguish myeloid sarcoma from malignant lymphoma. When the proliferating cells do not express the immunophenotype of a lymphoma, the pathologist should think about the diagnosis of myeloid sarcoma. The coexpression of some T-cell markers, for example, CD 2, CD 5, or CD 7, can cause difficulties in interpretation. TdT and CD 34 can be positive in both myeloid sarcoma and lymphoblastic proliferation. The clinical presentation and morphology must be considered. Typical markers of myeloid sarcoma must be studied systematically. Diagnosis can be particularly difficult in cases of transformation of chronic myeloid leukemia into a precursor T- or B-cell lymphoblastic lymphoma-leukemia. Note CD 56 is often expressed in myeloid sarcoma.

2. *Small round cell tumors.* This type of solid tumor, mainly observed in children, comprises neuroblastoma, rhabdomyosarcoma, Ewing's sarcoma, peripheral neuroectodermic tumor (PNET), and medulloblastoma. To distinguish among these diagnoses and myeloid sarcoma, various elements need to be considered: the clinical presentation, the site of the tumor, the morphology of the cells, a complete immunophenotype, and in some cases, the demonstration of specific genotypic lesions. The differential diagnosis based on morphology alone can be extremely difficult, in particular with Ewing's sarcoma and related tumors. In any patient for whom the morphology and the immunophenotype do not suggest a myeloid origin of the neoplastic cells, a search should be made for typical translocations (i.e., t(11-22) in Ewing's tumor) or transcripts of these translocations.

3. The diagnosis of *undifferentiated carcinoma* or *melanoma* can be eliminated by use of the same approach. *Malignant histiocytosis* made of very large cells with atypical nuclei is usually easy to identify. The diagnosis of *malignant mastocytosis* with atypical mast cells can be made only if metachromatic granules, even rare, can be demonstrated by Giemsa and toluidin blue staining or if the presence of tryptase is shown by immunohistochemistry [42].

4. *Extramedullary localizations of chronic myeloproliferative diseases* without blastic crises should be differentiated from myeloid sarcoma. These occur mainly in lymph node and are characterized by the accumulation of myelocytes and more mature myeloid cells in the nodal sinuses in the adjacent parenchyma and around vessels. They are sometimes associated with clusters of mature erythroblasts and mature megakaryocytes but do not have any CD 34-positive cells showing blast morphology. Patients with chronic myelomonocytic leukemia have frequent skin or mucosa localizations that can often be very difficult to distinguish from myeloid sarcoma. The absence of typical blast cells and of CD 34+ cells is the most important criterion for excluding the diagnosis of myeloid sarcoma.

5. *Some nonmalignant lesions* should also be included in the differential diagnosis.

Pseudotumors created by erythromyeloid metaplasia of adipose tissue occurring in the mediastinum and retroperitoneal areas are easy to recognize. The cells present include cells belonging to the three main hematopoietic series, which develop normally in the bone marrow, and are at different stages of maturation. There are no blasts, no CD 34-positive cells, and there is no maturation block. Pseudotumors occur in patients with severe hemolytic anemia or with extensive bone marrow destruction.

Some granulocytic proliferations with abundant immature myeloid cells, occurring in the spleen of patients treated by growth factors such as G-CSF, can be difficult to interpret, particularly if no clinical information is available [43].

Finally, plasmacytoid monocyte hyperplasia should be distinguished from nodal or cutaneous myeloid sarcoma. In such hyperplasia, there are nests or sheets of medium-sized cells with a round or ovoid nucleus, a small nucleolus, and an abundant and pale cytoplasm that stains faintly grey-blue with Giemsa stain, often with a plasmacytoid pattern of the cytoplasm. These cells express CD 68 and CD 4 but neither CD 34 nor B- or NK/T-cell associated antigens. They can occur in various reactive processes in lymph nodes (viral diseases, Castleman's disease) or in the skin (disseminated lupus erythematosus).

Pathogenesis

The cause of extramedullary tissue accumulation of myeloid blasts in tumors has not been identified; however, it may be due to abnormal expression of some adhesion molecules by the blasts. The expression of CD 56 (NCAM, Leu19) has been demonstrated on the blast membrane in some patients with myeloid sarcoma. This molecule may show homophilic binding to various tissues also expressing CD 56 (gastrointestinal tract, ovary, testis, skeletal muscle, nervous tissue). Blasts presenting the t(8;21) are often associated with CD 56 membrane expression and also with a high incidence of myeloid sarcoma developing during the initial phase of the acute leukemia or as a relapse after treatment.

Treatment and Disease Progression

Myeloid sarcoma should be treated in the same way as acute myeloblastic leukemia, even in cases of isolated tumors with no blood or bone marrow involvement. Radiotherapy has been proposed in association with chemotherapy for patients with massive tumors or for patients with spinal cord compression.

The progression of myeloid sarcoma in patients with acute leukemia has the same prognosis as the underlying leukemia. Patients with an acute myeloblastic leukemia associated with a t(8;21) and presenting myeloid sarcoma have a low rate of complete remission, and overall survival is poor [44].

Myeloid sarcoma in patients with chronic myeloproliferative disorder and myelodysplastic syndrome is equivalent to a blastic transformation of these diseases, and those patients often have a short survival.

After treatment of acute myeloblastic leukemia, myeloid sarcoma may develop as a relapse, even after bone marrow transplantation. Some chromosomal abnormalities such as t(8;21) seem to be predictive of disease relapse and should be screened for systematically by cytogenetic testing.

Acknowledgment

We thank Mr. Lionel Poursac for providing the photographs.

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