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Atypical Presentation of Myeloid Sarcoma Mimicking a Subcutaneous Hematoma as Complication of MDS and AML

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Abstract

Introduction: Less than 1% of patients with CML or acute leukemic transformations of other chronic disorders such as myeloproliferative neoplasms, particularly myelofibrosis, will present with and extramedullary presentation. Sites of isolated myeloid sarcoma include bone, periosteum, soft tissues, and lymph nodes, and less commonly the orbit, intestine, mediastinum, epidural region, uterus, and ovary.

Methods: We conducted a systematic review of cases reported in the literature since 2005 to 2013. We identified similar cases to the actual ones that presented to our practice. The patients were diagnosed with myeloid sarcoma with an atypical presentation as a hematoma. We described 2 case reports, both have an atypical presentation of myeloid sarcoma mimicking a hematoma in different parts of the body.

Results: We found in the literature a total of 4 cases with myeloid sarcoma with an atypical presentation as a hematoma. About, 3 of them (n=3, 75%) reported subdural hematomas. Only 1 case (n=1, 25%) reported an ocular involvement/retro-orbital hematoma. We present 2 cases that presented to our practice with a diagnosis of myeloid sarcoma and later on complicated as a subcutaneous hematoma.

Discussion: We analyzed the fact that myeloid sarcoma has a very unusual presentation as a hematoma. The review of the literature supports this fact. However this should make as aware that even though this is an unusual and unique presentation we should think about it and always test the biopsy sample for IHC and flow cytometry in order to later on diagnose and treat

Keywords: Myeloid sarcoma; Chloroma; AML; CML; Myeloproliferative disorders; Hematoma

Introduction

About less than 1% of patients with CML or acute leukemic transformations of other chronic disorders such as myeloproliferative neoplasms, particularly myelofibrosis, will present with and extramedullary presentation. These presentations are known as myeloid sarcoma, also called granulocytic sarcoma, myeloblastoma, or chloroma. When found in association with blood or bone marrow involvement, it occurs most commonly as either cutaneous or gingival infiltration by leukemic cells [1]. However, sites of isolated myeloid sarcoma include bone, periosteum, soft tissues, and lymph nodes, and less commonly the orbit, intestine, mediastinum, epidural region, uterus, and ovary [2]. Myeloid sarcoma should be suspected in the presence of a "small round blue cell tumor," and should be more seriously suspected if eosinophilic myelocytes are seen on hematoxylin and eosin-stained biopsies [3]. We will present 2 cases and a review of the literature of an atypical presentation of a myeloid sarcoma mimicking a hematoma.

Methods

We conducted a systematic review of cases reported in the literature since 2005 to 2013. The patients were diagnosed with myeloid sarcoma with an atypical presentation as a hematoma. Only patients with prior diagnosis of "myelogenous leukemia", either "acute" or "chronic", "chloroma" and "myeloid sarcoma", were included in this study. Patients with "hematoma" were included. There were no cases identified of "subcutaneous" "hematoma" previously described in the literature to the best of our knowledge. Baseline demographic, clinical and laboratory data were collected. We identified a total of 4 cases reports with similar features to the ones that we encountered in our practice. We describe 2 case reports, both have an atypical presentation of myeloid sarcoma mimicking a hematoma in different parts of the body. These presentations are very unique and never described before in the literature.

Results

Review of the literature

We conducted a systematic review of the literature for the presentation of myeloid sarcoma as a hematoma. We found a total of 4

cases. A total of 3 (75%, n=3) reported subdural hematomas. Only 1 of the cases (25%, n=1) reported an ocular involvement [4-7]. There are not much descriptions of chloromas or myeloid sarcomas presentations in the literature as hematomas. This unique presentation makes us aware of this atypical phenomenon.

Baseline Characteristics

In this review, we included a total of 4 cases of myeloid sarcoma mimicking a hematoma reported in the literature from 2005-2013. The median age of diagnosis was 53.5 years and the majority of the patients were male (75%, n=3) (Table 1). The clinical presentation at diagnosis was consistent with headaches (75% n=3). None of them were asymptomatic at the time of diagnosis. The most frequent site of hematomas was subdural (75%, n=3) followed by orbital (25%, n=1).

Characteristic	N (%)
Age at initiation of therapy	
<65 years	3 (75.0%)
≥ 65 years	1 (25%)
Sex	
Male	3 (75.0%)
Female	1 (25.0%)
Race	
Caucasian	2 (50.0 %)
African-American	2 (50.0%)
Chloroma locations	
Subcutaneous	0 (0.00%)
Brain	3 (75%)
Eye	1 (25%)
Prior treatment	
TKIs	1 (25.0%)
Hydroxyurea	1 (25.0%)
Cyclophosphamide + BMT	1 (25.0%)
Unclear	1 (25.0%)

Table 1: Baseline characteristics of patients included in the analysis of cases reported in the literature.

Case 1

A 71-year-old female with 1-year history of MDS, complicated by persistent pancytopenia, recurrent neutropenic fever and AML transformation, treated with decitabine. She presented to the office complaining of spontaneous subcutaneous mass in her right upper arm, which occurred over a 48h period. At the time, CT of chest/abdomen confirmed a 2.4 cm subcutaneous mass, without any significant lymphadenopathy or active pulmonary disease. Excisional biopsy revealed organized blood clot with dispersed atypical cells of unclear origin by immunohistochemistry (IHC). During a one-month period, the mass became gradually enlarged, edematous, and erythematous. Physical examination on admission was significant for fever (100.3 F) and right upper arm mass with tenderness, warmth, and erythema. Laboratory results showed Hg of 8 g/dL, WBC of 500 cells/uL (neutrophil 42%, lymphocyte 50%, blast 6%, absolute neutrophil count 210 cells/uL), platelets of 43,000/uL, INR of 1.14, and PTT of 41.5. Multifocal nodules with small ground glass opacities throughout lung parenchyma were detected on repeated chest CT. They were subsequently found to be due to invasive pulmonary aspergillosis, in which voriconazole was initiated. This time, incisional biopsy with debridement of the mass were done. Pathology revealed dermal infiltration of blast cells with myeloid cytoplasmic granulation; IHC revealed positive CD15 and myeloperoxidase, occasionally positive CD34, negative CD34. Small lymphoid cells also stained for leukocyte common antigen (LCA). The findings supported the myeloid origin of these cells. (Figure 1).

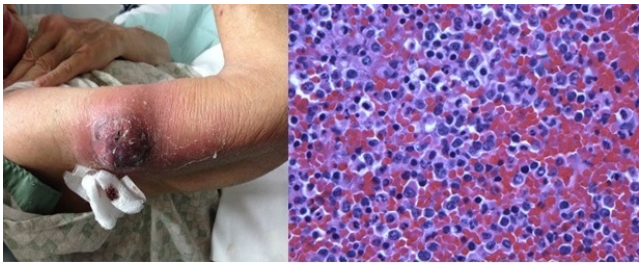

Figure 1: Lesion on right arm, consistent with hematoma, MWMC. Blasts, blue myelocytes and red blood cells on biopsy obtained from hematoma above, Hematoxylin and eosin-stained biopsy Dr. Saint Aufranc, MWMC.

Case 2

A 69-year-old male with diagnosis of Myelodysplastic Proliferative Syndrome with decreased erythropoietin, transfusion dependent, refractory anemia with excessive blasts, CML complicated by AML. He was on treatment with hydroxyurea, later on stopped and switched to chemo despite multiple comorbidities. Was on combined chemo with azacitidine and vidaza. Patient was diagnosed of MDS and MPS through bone marrow biopsy. No transformation to leukemia at the time. Later on the patient developed hypoxemia which lead to multiple hospitalizations. He was found to have an abnormality in the lung, which led to further evaluations with a CAT scan showing multiple bilateral lung nodules and cavitary lesions in the left lung. He has had a biopsy of the right lung lesion, confirming aspergillosis. He was started on voriconazole for several weeks. However, worsening of SOB lead to further evaluation showing blasts increasement on the peripheral blood consistent with Myelodysplastic syndrome with excess blasts with transformation into acute myelogenous leukemia with atypical presentation due to infiltration of the lymph nodes with myelosarcoma cells with no peripheral blood indicative of active transformation. Patient was started on decitabine and intermittent courses of hydroxyurea when the leukocytosis worsens due to AML. About 1 year after initial MDS diagnosis, patient was noted to have an enlarged bi-axillary lymph nodal enlargement and pleural effusion. He had a biopsy of the lymph node and also pleural fluid cytology analyzed. Lymph node biopsy from the left axillary lymph node showed evidence of a myeloid neoplasia with myeloid sarcoma. His pleural fluid analysis

also showed inflammatory cells and also mononuclear cells indicative of myelodysplastic proliferative cell etiology. Immediately after this lymph node transformation a rapid growing mass in the left leg started to show up. This mass was an ovoid, flattened deep brown red segment of tissue. Biopsy showed a blood clot embedded with irregular aggregates with degenerated small to intermediate cells. IHC revealed a positive staining of a subset of degenerated mononucleated cells for CD34 and prominent staining for myeloperoxidase. Scattered cells are positive for CD117. Background small lymphocytes stain for LCA with a predominance of T-Cells (CD3) and scattered B-cells (CD-20, PAX-5). These current findings were consistent with myeloid sarcoma. Patient received surgical excision in top of his chemo with decitabine, responded appropriately but passed after 1 month of diagnosis due to his other comorbidities (Figure 2).

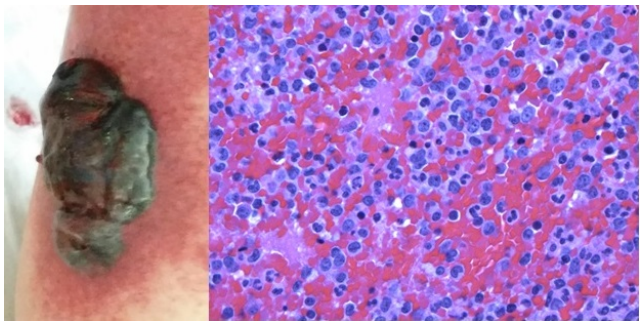

Figure 2: Blasts, blue myelocytes and red blood cells on biopsy obtained from hematoma of the left leg visualized on the right side of this image. Hematoxylin and eosin-stained biopsy Dr. Saint Aufranc, MWMC.

Discussion

When we started to evaluated these cases, we knew were unique due to the fact that there is very limited information in the literature to describe this entity. The review of the literature supports this fact. We found a total of 4 cases in the literature with the features of a myeloid sarcoma mimicking a hematoma. In this review we emphasize that to the best of our knowledge, these are the only two cases so far described as a myeloid sarcoma or chloroma mimicking a subcutaneous hematoma. These two cases had in common to appear spontaneously and grow really fast. Not following at all the usual natural history of a regular hematoma.

However this should make us aware that even though this is an unusual and unique presentation, we should think about it and always test the biopsy sample for IHC and flow cytometry in order to obtain an accurate diagnosis and be able to treat accordingly without delay. Both of our patients appeared to have a rapid deadly progression of their disease after this presentation. The early identification of this entity might be able to improve survival in these patients. The power of this study is too small, granting further studies of myeloid sarcoma in this scenario.

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