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Case Report

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De novo Extramedullary myeloid tumors and chronic myeloid leukemia at chronic phase on a Congolese patient

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ABSTRACT

Extramedullary myeloid skin had tumor is a rare entity. It usually appears during the life time of a blastic myeloid disorder. We report a rare case of a 62 years old female patient who developed extramedullary myeloid skin tumors which had preceded chronic myeloid leukemia at chronic phase.

BACKGROUND

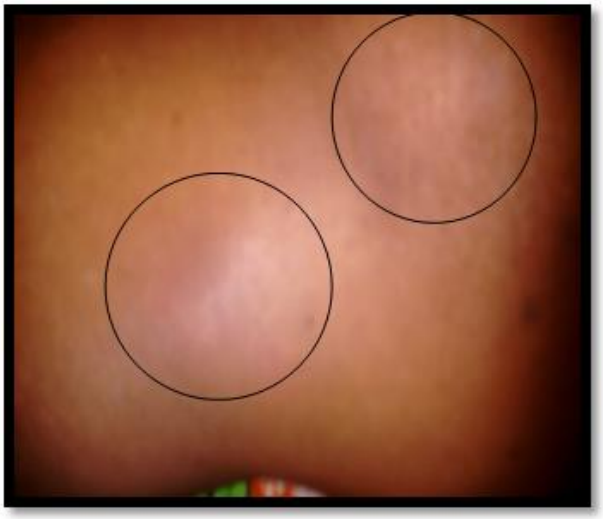
Extramedullary myeloid tumor (EMT) or granulocytic sarcoma is a rare extramedullary tumor. It refers to a skin infiltration of myeloid malignancy. Depending on the chronology of the skin lesion, Benet et al distinguish three groups of EMT: de novo, consequent and subsequent [1]. In the de novo group, the skin lesion precedes any underlying myeloid disorders suspected or diagnosed. In the consequent group, the skin is found while the myeloid disorder is diagnosed. Finally in the subsequent group, the skin lesion appears during the life time of the myeloid disorder.Most of EMTs are associated with acute myeloid leukemia or refractory anemia, chronic myelomonocytic leukemia and chronic myeloid leukemia at their blastic phase. Therefore, diagnostic of EMT can be very challenging outside the context of blastic malignancies. A few cases of EMT associated with chronic myeloid leukemia (CML) at chronic phase have been reported, however most of them were belonging to the consequent or subsequent groups [1, 2, 3]. We describe in this paper the case of an EMT for which the diagnosis was not readily made because in one hand it was preceding an unknown myeloid malignancy (CML) which on another hand was at the chronic phase of its development.

CASE PRESENTATION

A 62-year-old women with no previous medical history presented to dermatology outpatients department for 6 months painful skin lesions of arms and thighs. She denied fever, loss of weight or fatigue. At the initial clinical examination, the patient that was afebrile and hemodynamically stable. The skin examination

showed painful reddish subcutaneous nodular lesions, pruriginous, 2-3 cm in diameter from the external surface of the arm and posterior aspect of the right thigh (Figure 1). On the abdominal examination they were not lymphadenopathy detected, splenomegaly nor hepatomegaly. The cell blood count showed a leukocytosis at 9.8 giga/L with differential of 85% neutrophil polynuclears, 3% neutrophil metamyelocytes, and 11% lymphocytes, normocytic anemia, normochromic at 11 g/dL and platelet count at 302 giga/L.

Figure 1: Two Subcutaneous reddish nodular lesions



A biopsy resection of the cutaneous lesions was carried out and sent in Avicenne Hospital in France for investigation. Histological analysis showed hematopoietic extramedullary proliferation of granular neutrophils and eosinophils at all stages of maturation and numerous megakaryocytes without blast cells. The immunehistochemical study was positive for myeloperoxidases (50%), the CD15 antigen was positive (60%). Because they were not underlying myeloid disorders known or suspected, myeloid markers as CD 68 and CD 33 were not studied. However since EMT was one of the differentials diagnosed considered by the pathologist, cytogenetic studies to look for Philadelphia chromosome were recommended.

Unfortunately, because the patient could not afford to pay for the cytogenetic studies, she was lost of follow up.

The patient showed up 4 months later because the size of her skin lesions increased and she was complaining about abdominal pain. Besides the skin tumors, we found a splenomegaly of 10 cm high on the mid-clavicular line below the last palpable rib without lymphadenopathy nor hepatomegaly.

The second cell blood count showed a leukocytosis at 48.8 giga/L with differential of 82% neutrophil polynuclears, 11% eosinophil polynuclears, 2% neutrophil metamyelocytes, 1% neutrophilic myelocytes, 1% eosinophilic myelocytes, 2% blasts and 1% lymphocytes, normocytic anemia, normochromic at 10.1 g/dL and platelet count at 502 giga/L.

Bone marrow aspiration showed myeloid hyperplasia. Cytogenetic studies performed on the bone marrow aspiration revealed positive Philadelphia chromosome. Polymerase Chain Reaction study identified BCR-ABL (e13a2 and e14a2). The sokal score was 1.33.

Based on the clinical, histological and biological findings we retrospectively diagnosed skin lesions as EMTs that were belonging to the De novo group and high risk chronic myeloid leukemia at chronic phase.

Imatinib was initiated at the dosage of 400 mg per day orally associated with Hydroxyurea 2000mg daily. A complete hematological response was achieved under treatment in 18 days. Skin tumors and splenomegaly disappeared. The patient has been followed up for 6 months and she is doing well.

DISCUSSION

The extramedullary myeloid tumor (EMT), also called granulocytic sarcoma, formerly chloroma, is a rare malignant haemopathy of extramedullary localization composed of more or less differentiated immature cells [4].

The incidence in the United States is 2 cases / million inhabitants in adults [5]. The development of EMT during AML is poor prognosis [1].

EMT is mainly described in two major groups of malignant blood disorders: acute myeloid leukemia (AML) or secondary to chronic myeloid leukemia, myelodysplastic syndrome or myelomonocytic leukemia in transformation phase [4].

When EMT is the initial manifestation of any underlying or diagnosed myeloid malignancies, it

belongs to the called De novo EMT's group and is also designed as aleukemic lesion [1]. The diagnostic diagnosis of the tumor in this context is extremely difficult and can be confused with lymphoma, carcinoma or sarcoma [1, 4]. De novo EMT is extremely rare and account for 7% of all aleukemic lesions [1]. It can occur in all part of the body, but skin localization is the most common presentation [3].

Histological characteristics of EMT are not specific to underlying hematological malignancies [1, 4].Thus, only the immunohistochemical characteristics expressed by the tumor confirm the identity of the tumor. In myeloid malignancies characteristics of the tumor are defined by the presence of markers as the myeloperoxidase, antigens CD13, 33, 68 and CD 117 [1, 3].

Even though the patient was lost of follow, two factors disoriented our diagnosis. First, De novo characteristic of the tumor did oriented us to search for antigens specific myeloid malignancies. Second, the chronic development phase of the CML did not lead us to think about extramedullary manifestation of the CML because it was at its chronic phase development. Indeed, EMT associated with chronic myeloid leukemia at its chronic phase is unusual [2, 6]. Bone marrow examination and clinical findings were the one that actually lead us to the diagnosis of chronic phase CML. Diagnosis of EMT was done retrospectively and confirmed by disappearance of skin lesions under imatinib.

CONCLUSION

This observation reminds us that EMT can precede, be consequent or subsequent to any myeloid disorders at their all phases of development. It is essential to the clinician to be familiar with the various clinical features of the chronic myeloid leukemia which is the most common chronic leukemia in the Congo.

CONFLICT OF INTERESTS

The authors declare that there is no conflict of interests regarding the publication of this paper.

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