Cancer Therapy & Oncology
International Journal
ISSN: 2473-554X

Juniper Publishers
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Case Report
Volume 7 Issue 4 - October 2017
DOI: 10.19080/CTOIJ.2017.07.555716

Canc Therapy & Oncol Int J

Multiple Skin Granulocytic Sarcomas (Chloroma) - as an Initial Presentation of Cml in a Teenager- A Cyto-Hematologic Correlation

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Submission: October 03, 2017; **Published:** October 23, 2017

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Abstract

Granulocytic sarcoma (chloroma), as an initial presentation of CML (chronic phase) in a teenager is a very unusual and rare presentation. Granulocytic sarcomas are rare malignant extra-medullary tumour of primitive granulocytic cells that usually occurs in acute myeloid leukemia or blast phase of chronic myeloid leukemia (CML). We report a case of CML in chronic phase, presenting initially as multiple subcutaneous granulocytic sarcoma in a teenager, who landed in the orthopedic department, for complaints of pain and multiple swelling in both the legs for a short duration.

Introduction

The name Chloroma (Granulocytic sarcoma), is derived from the greek word chloros (green), due to the enzymatic action of myeloperoxidase in the tumour cells, which imparts a greenish color to the tumour in most of the cases. They can arise de novo 5 or can be associated with acute myeloid leukemia (AML), chronic myeloid leukemia (CML) and other myeloproliferative or myelodysplastic conditions [1,2]. Rappaport6 suggested the term granulocytic sarcoma considering the association of these tumors to leukemia. It can involve any anatomical site but most often involves the lymph-node, bone, periosteum, soft tissue and skin. It can be a great challenge to diagnose this case on cytologic specimen, if not suspected clinically and without any hematological investigations. Cytomorphology of chloroma is sparse [1-3]. Granulocytic sarcoma is a great mimicker and should be included in the differential diagnosis of aspirates from skin, soft tissues or lymph nodes.

Case Presentation

A sixteen year old boy landed in the Orthopedic O.P.D. with complaints of pain and swelling in both the legs since 15 days, with associated history of loss of weight since few months. He had localized pain over the multiple swellings on the anterior aspect of both the legs of 15 days duration. Examination revealed multiple firm, nodular swellings, the largest measuring 4x3.5 cm and the smallest measuring 2x1 cm. X-ray of both the legs revealed no bony or periosteal involvement. Ultrasound abdomen revealed a mild splenomegaly (6cm from left sub costal line). CRP (3mg/l) and Uric acid (7.8 mg/dl) were within normal limits for the patients age.

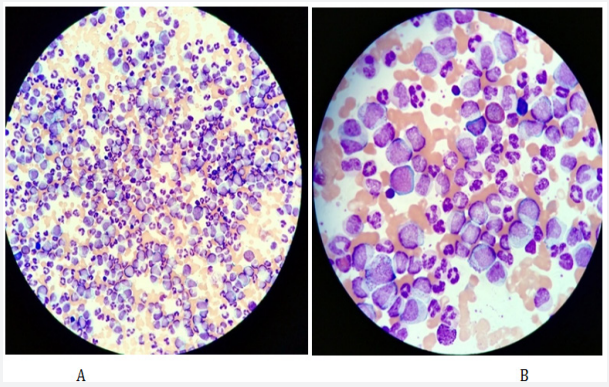

Figure 1
A. Leishman Stain-10x
B. Leishman Stain-40x

On hematological investigations, hemoglobin was 8.8 g/dl with low hematocrit (23.8%), very high total leucocyte count(419.9 x 10^3/µl), platelet count was mildly increased (519x10^3/mm^3), with the following differential counts: Blasts-2%, Promyelocyte-15%, Myelocyte-18%, Metamyelocyte-11%, Neutrophils-46%, Lymphocytes-2%, Eosinphils-1% and Basophilia -5%. Features were suggestive of

Chronic Myeloid Leukemia (CML)- in chronic phase. Leucocyte Alkaline Phosphatase Score (LAP) was 8 (low score). Fine needle aspiration (swelling on both the legs) and RT-PCR for BCR-ABL translocation study from peripheral blood (EDTA sample) was advised (Figure 1A & 1B).

(Both the picture shows marked leucocytosis with prominent basophilia, marked left shift with myloblasts).

Cytology smears studied from the aspirate of the subcutaneous swelling on both the legs, showed cellular smears composed of promyelocyte, myelocyte, metamyelocyte, neutrophils and eosinophils scattered in a discohesive pattern. Scattered eosinophilic metamyelocytes are seen. Occasional blasts are noted. The above cytological features were suggestive of Granulocytic Sarcoma- pretibial skin.

(The above cytology smears (Figures 2A-2C) show, high cellularity composed of promyelocyte, myelocyte, metamyelocyte, neutrophils, eosinophils and eosinophilic metamyelocytes are seen. Occasional blasts are noted.) BCR/ABL1 Translocation Assay (Quantitative) by Minor Groove Binder Real Time RT-PCR showed BCR/ABL 1 t (9:22) (q34; q11)- BCR-ABL/ABL1 transcript ratio (%) is 100%.This confirms our diagnosis of Granulocytic sarcoma with CML in chronic phase.

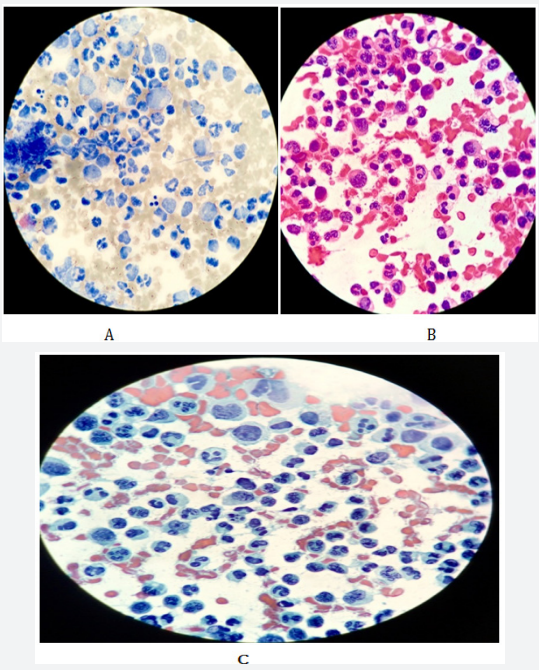

Figure 2
A. MGG-40X
B. H&E-40X
C. PAP-40X

Discussion

Granulocytic sarcoma was first described in 1811 by Burns and was originally called "Choloroma" by King in 1853. It is a rare solid tumour composed of immature myeloid cells and can appear at any age any site. It is known by several other names, such as Myeloid Sarcoma, Myeloblastoma, Chloromyeloma, Chloromyelosarcoma, and Chloroma. W.H.O has classified granulocytic sarcoma into 3 main types, depending upon the degree of maturation of the cells of myeloid series. They are:

i. Blastic- composed of mainly myeloblasts.

ii. Immature- composed of myeloblasts and promyelocytes.

iii. Differentiated- predominantly promyelocytes and more mature myeloid cells.

iv. The less common type - Monoblastic sarcoma [4-6].

Occurrence of chloroma in patients with myeloid leukemia indicates warning signs of acute myelogenous leukemia or onset of accelerated disease or blast crisis of CML [7,8]. The tumour is capable of local soft tissue invasion.

The differential diagnosis of granulocytic sarcoma can be:

a. Extra medullary hematopoiesis.

b. Infectious process.

In our case, this boy presented with a short history of multiple swellings in both the legs, which was diagnosed as granulocytic sarcoma of skin on FNAC, in conjuction with the peripheral blood picture of CML in chronic phase, which was later confirmed by RT-PCR. It is an extremely rare manifestation. The case presented here showed suggestive evidence linking the manifestation to the disease. These type of cases if not taken in toto, can pose quiet a diagnostic dilemma to the orthopedician and the cyto-pathologist alike. A wholesome approach is therefore needed in these cases for diagnosis and treatment. Therapies include local radiation, systemic chemotherapy, immunotherapy, surgery, hematopoietic stem cell transplantation, and donor lymphocyte infusion and combination therapy.

Conclusion

Granulocytic Sarcoma requires early detection because a local relapse could be alleviated by local therapy (radiation). We report this case due to its rarity and unusual presentation in a teenager. Granulocytic sarcoma (chloroma) should be kept as a differential diagnosis in the mind of orthopedic surgeons for early detection, and rigorous diagnostic workup.

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How to cite this article: Sreeya D, Mamina B, Dipika M . Multiple Skin Granulocytic Sarcomas (Chloroma) - as an Initial Presentation of Cml in a Teenager- A Cyto-Hematologic Correlation. Canc Therapy & Oncol Int J. 2017; 7(4): 555716. DOI:10.19080/CTOIJ.2017.07.555716.

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