International Journal of Research in Medical Sciences
Bansod YV et al. Int J Res Med Sci. 2013 Aug;1(3):291-293
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: 10.5455/2320-6012.ijrms20130828

Case Report

Ascites in CML: a rare extramedullary manifestation

Y. V. Bansod, Sandeep V. Kharkar*, Arshish Raut, Piyush Choudalwar

Department of Medicine, Government Medical College, Nagpur, Maharashtra, India

Received: 10 May 2013
Revised: 30 May 2013
Accepted: 31 May 2013

***Correspondence:**
Dr. Sandeep V. Kharkar,
E-mail: drsandeep2k@yahoo.com

ABSTRACT

Ascites in chronic myeloid leukemia (CML) as an extramedullary manifestation is rarely reported in the literature. A young 23 year old female of chronic myeloid leukemia presenting with extramedullary disease as massive ascites. All stages of granulocytes and a few blasts similar to peripheral blood smear was present in ascitic fluid. Based on clinical symptoms and signs, bone marrow examination, ascitic fluid cytology and ultrasonography of abdomen she was diagnosed as a case of Chronic myeloid leukemia in chronic phase with extramedullary disease as massive ascites. After starting treatment with Imatinib mesylate favourable response was observed.

Keywords: Ascites, Chronic myeloid leukemia, Extramedullary, Imatinib mesylate

INTRODUCTION

Chronic myeloid leukaemia (CML) is a clonal myeloproliferative stem cell disorder, characterized by expansion of myeloid, erythroid, and megakaryocytic cells. with an incidence of 1-2 cases per 100,000 populations.

The clinical course of (CML) is characterized by two different clinical phases the initial chronic phase, and the subsequent acute phase or blast crisis. After 2- to 4-year chronic phase, the disease accelerates with an increase in the percentage of blast cells in the peripheral blood and bone marrow.[2]

Some proportion of patients with CML develops extramedullary disease caused by the infiltration of blast cells this condition is called extramedullary blast crisis. The incidence of extramedullary blast crisis is reportedly 7 to 17 percent in patients with CML.[2]

We hereby report a patient of CML in chronic phase who developed extramedullary blast crisis in the form of massive ascites with no increase in blast cells in the bone marrow, and who was put on Imatinib mesylate with a favourable response.

CASE REPORT

A 23 years Indian female residing in rural area with low socioeconomic status farm worker by occupation presented with generalised weakness and easy fatigability of three months duration, pain and distension of abdomen since 15 days and difficulty in breathing since 3 days .She had no fever, cough, jaundice, decreased urine output, chest pain , palpitations or bleeding tendency. No family history significant. On physical examination, patient was pale, emaciated, afebrile, pulse 90/min, blood pressure was 110/70 mmHg, respiratory rate of 24/min and SpO2 was 92% on room air, JVP was normal, no cyanosis, oedema feet, lymphadenopathy, sternal tenderness or any signs of liver cell failure. Abdominal examination revealed tense ascites with massive splenomegaly. Normal peristaltic sounds were audible. Rest of the systemic examination was within normal limits.

Laboratory investigations revealed HB: 7.0 gm%, PS: TLC > 2,00,000 /mm^3, DLC: N -58%, L-7%, M-7%, E-3%, B-3%, promyelocyte 2%, myelocyte 10%, metamyelocyte 4%, blast 6%, platelets - 5,08,000/ mm^3. Bone marrow examination showed hypercellular marrow with all stages of granulocytic hyperplasia and 6% blasts. Ascitic fluid analysis revealed glucose 109 mg/dL, protein -5.2 g/dL (serum protein 6.6 g/dL), albumin 2.9g/dL (serum albumin 3.2 g/dL).Ascitic fluid microscopy revealed WBC count 2,390 /mm^3 (N-57%,L-6%,E-2%,B-3%, promyelocyte 3%, myelocyte 17%, metamyelocyte 7%, blast 5%). The ascitic fluid was negative for Gram stain and acid fast bacilli. Ultrasound examination of abdomen revealed gross ascites with spleen measuring 19.3 cms. Liver and portal vein were within normal limits.

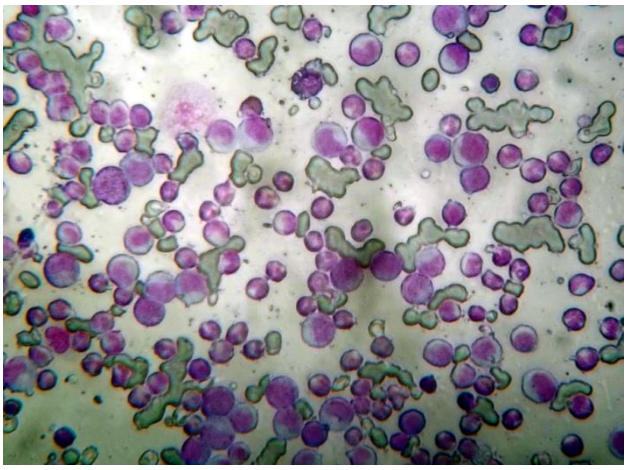

Figure 1: Peripheral smear shows a full spectrum of cells in the granulocyte series, ranging from blast forms to mature neutrophils, with intermediate myelocytes and neutrophils.

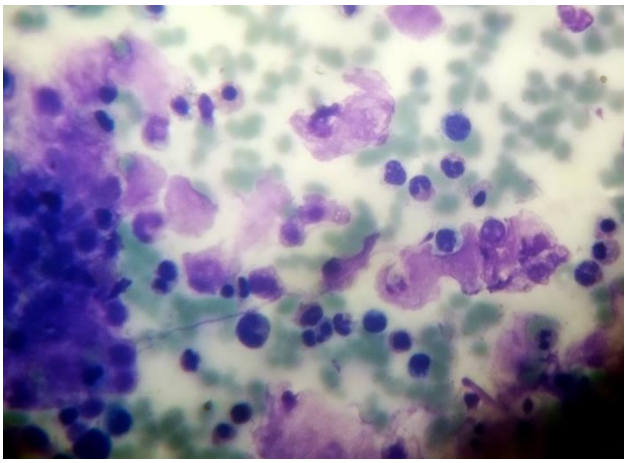

Figure 2: Smear from ascitic fluid showed a dense infiltrate of immature granulocytic precursors along with a few eosinophils and basophiles in the background of RBCs.

Patient's coagulation profile, liver and kidney function tests was normal. Her relative risk scores like sokal score was 0.65 (low) and hasford score was 220.67 (low). Patient was diagnosed as a case of CML in early chronic phase with extramedullary disease as massive ascites with low relative risk scores. Patient was started on tablet Imatinib 400 mg OD and a favourable response in the form of regression of spleen and ascites, her blood counts came to the normal with normal Hb and TLC counts with no blasts in peripheral smear at the end of 4 months of treatment.

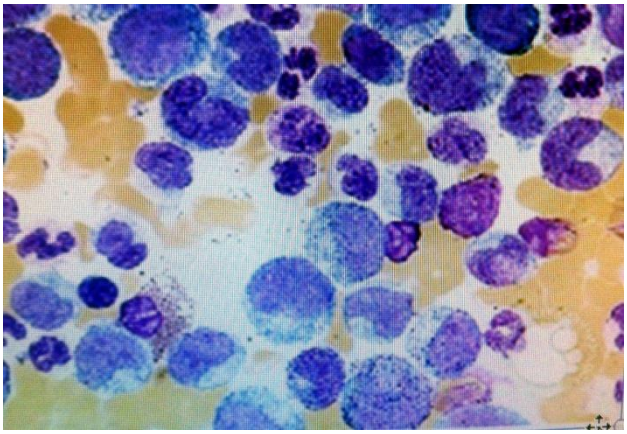

Figure 3: Smear of marrow aspirate showing an increased number of granulocytes in all stages of development and blasts (Wright-Giemsa stain).

DISCUSSION

Although extramedullary blast crisis is generally accompanied by increasing blasts in the bone marrow or peripheral blood, it may precede medullary disease progression.[2] Extramedullary blast crisis is the first manifestation of accelerated phase in approximately 10 percent of patients with CML.[3]

Lymph nodes, serosal surfaces, skin and soft tissue, breast, gastrointestinal or genitourinary tract, bone and central nervous system are among the principal areas involved.[3] Case reports of pleural effusion,[4] pericardial effusion[9] and cardiac tamponade[8] in patients with CML have been reported. Ascites in chronic myeloid leukaemia (CML) is rarely reported.

Umesh das *et al and* Hyun woo kim *et al* have proposed four mechanisms for the development of effusions in patients with CML.[4,5]

The first mechanism is leukemic infiltration into the visceral cavity.[5]

The second mechanism is reaction secondary to bleeding into the cavities that may cause effusions and ascites. Predisposing factor such as leukostasis and platelet dysfunction may have a role in hemorrhagic effusion. If

this is true, the ratio of red blood cells to nucleated cells in the blood and ascitic fluid should be similar.

A third possible mechanism underlying effusions is extramedullary haematopoiesis.

A fourth mechanism causing effusion is non malignant causes, such as infection.

Therefore, the possibility of presence of necrotic debris and identification of microorganisms may suggest an infectious process.[5]

Generally the median time from diagnosis of extramedullary blast crisis to marrow blast crisis is 4 months, and the median survival is 6 months.[6] Extramedullary disease in CML is considered as an indicator of poor prognosis.[6] Imatinib mesylate, a competitive inhibitor of the Bcr-Abl tyrosine kinase, is highly effective in treating chronic phase CML, but less effective against CML in its advanced phases. However, the efficacy of Imatinib mesylate for extramedullary blast crisis of CML has yet to be fully elucidated.[2]

In conclusion, patients with CML should be considered at risk for development of extramedullary manifestations of blast crisis while the bone marrow remains in the chronic phase.[4] Extramedullary blast crisis of CML can occur at anytime and circulating stem cells can be seen anywhere.[4] In CML patient with ascites, bleeding into peritoneal cavity and extramedullary haematopoiesis should first be considered.

REFERENCES

1. E.L. Morris and J.P. Dutcher (2005). Blastic Phase of Chronic Myelogenous Leukaemia. Clinical Advances in Haematology & Oncology Volume 3.
2. Naito K, Mori T, Miyazaki K, Tsukada Y, Ikeda Y, Okamoto S (2003). Successful treatment of extramedullary blast crisis of chronic myelogenous leukemia with imatinib mesylate (STI571). Intern Med.42(8):740-2.
3. K. Kaushansky, M. Lichtman , E. Beutler, T. Kipps, J. Prchal , U. Seligsohn (2010) Williams Hematology, Eighth Edition, edited by K. Kaushansky, E. Beutler, U. Seligsohn (The McGraw-Hill Companies, Inc).
4. U. Das, V. K. Gupta, T. Nyandak, P.Yadav, SC Sharma,D. Srivastava, M.Kaushal.(2010) Pleural Involvement in Chronic Myelocytic Leukaemia –an Extra-medullary Blast Crisis. JIACM 11(4): 326-9.
5. H. W. Kim, S. S. Lee,M.H. Ryu, J. L. Lee,H. M. Chang,T.W. Kim,H.S. Chi*, W. K. Kim,J.S.Lee, Y.K. Kang(2006). A Case of Leukemic Pleural Infiltration in Atypical Chronic Myeloid Leukemia. J Korean Med Sci; 21: 936-9.
6. Sacchi S, Temperani P, Selleri L, Zucchini P, Morselli S, Vecchi A,Longo R, Torelli G, Emilia G, Torelli U(1990). Extramedullary pleural blast crisis in chronic myelogenous leukemia: cytogenetic and molecular study. Acta Haematol; 83: 198-202.
7. Aleem A, Siddiqui N(2005). Chronic myeloid leukemia presenting with extramedullary disease as massive ascites responding to imatinib mesylate. Leuk Lymphoma.46(7):1097-9.
8. S Sobolewski, N Sreeharan, and S M Rajah (1979). Successful management of cardiac tamponade in two cases of leukaemia. Br Med J. November 10; 2(6199): 1190.
9. D.Hammouda, M.Shaheen, R.Nassar(2011). Imatinib (Gleevec) Induced Worsening of Pericardial Effusion in CML. Fırat Tıp Dergisi; 16(2): 100-1.
10. Sokal JE et al, Blood 1984; 63: 789-99.
11. Hasford J et al, JNTL Cancer Inst 1998; 90: 850-8.

DOI: 10.5455/2320-6012.ijrms20130828
Cite this article as: Bansod YV, Kharkar SV, Raut A, Choudalwar P. Ascites in CML: a rare extramedullary manifestation. Int J Res Med Sci 2013;1:291-3.