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Case Report

Extramedullary hematopoiesis in ovary: a rare presentation of CML

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ABSTRACT

Extramedullary hematopoiesis (EMH) is a rare phenomenon, and represents infiltration and proliferation of myeloid, erythroid, and megakaryocytic cells in non-bone marrow sites. Extramedullary hematopoiesis (EMH) is normal during fetal life, but after birth, the presence of EMH is considered to be abnormal. EMH has been associated with CML (chronic myeloid leukaemia). Lymph node is the most common site of involvement, other sites being the abdomen (liver, spleen) and thorax (bone, mediastinum) but very rarely in the gynecological tract. Here authors report a case of a 20-year young female, a known case of CML who presented with abdominal pain with hemoperitoneum, with negative urine pregnancy test. Initially hemorrhagic corpus luteal cyst was thought to be the cause of hemoperitoneum and patient was taken for surgery wherein histopathology confirmed the diagnosis of extra medullary hematopoiesis of ovary. Women with CML when presents with hemoperitoneum, extramedullary hematopoiesis should be a differential diagnosis apart from rupture ectopic and other causes of spontaneous hemoperitoneum. Patient was started on imatinib after diagnosis and is doing well.

Keywords: Chronic myeloid leukaemia, Corpus luteal cyst, Extramedullary hematopoisis, Haemoperitoneum

INTRODUCTION

Chronic myelogenous leukemia (CML) is a myeloproliferative neoplasm characterized by abnormal hyperplasia of granulocytes, which is secondary to the reciprocal translocation of chromosomes 9 and 22 and the consequent function of the BCR/ABL fusion gene.¹

CML accounts for approximately 14% of leukemia's worldwide with an incidence of 1-2 per 100,000 persons.^{2,3}

In CML, multiple blood stem cells become a type of white blood cell called granulocytes. These granulocytes are abnormal and do not become healthy white blood cells. They are also called leukemia cells. The leukemia

cells can build up in the blood and bone marrow so there is less room for healthy white blood cells, red blood cells, and platelets. When this happens, infection, anemia, or easy bleeding may occur.

CML is a primary disease of bone marrow and is classically divided into three phases (based on blast count and other parameters): chronic, accelerated, and blast crisis.

Most patients present in the chronic phase. Transition to the accelerated phase may be subtle or may be characterized by worsening anemia, splenomegaly and organ infiltration by hematopoietic elements.⁴ CML in the accelerated phase doesn't respond as well to treatment as CML in the chronic phase.

Extramedullary hematopoiesis (EMH) is the production of elements of erythroid and myeloid series at ectopic sites; when concomitant with agnogenic myeloid metaplasia, it is invariably seen in advanced disease.

This hematopoietic infiltration is very rarely seen in the gynecological tract.^{5,6} The most common sites for EMH are the liver, the spleen, and paraspinal regions of the thorax; however, it has been reported to involve virtually any organ or tissue.

Authors herein present a rare case of extramedullary hematopoietic infiltration in ovaries in a young female who was a known case of CML, who presented with abdominal pain with hemoperitoneum.

CASE REPORT

A 20 year married female presented to our emergency department with complain of severe pain in abdomen along with vomiting. Her last menstrual period was 20 days back.

She was diagnosed as CML at age of 17 but was not on treatment for last 4 months. On examination she was in early hypovolemic shock (pulse of 140/minute, BP of 100/60mmHg) with moderate abdominal distention and tenderness. Pelvic examination revealed bulky uterus with posterior fornix boggy. The urine pregnancy test was negative.

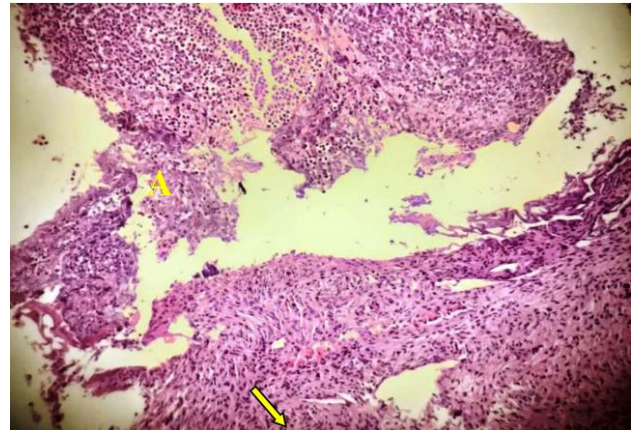
Investigations showed haemoglobin of 5.9g/dl, TLC- $4.49 \times 10^5/\text{mm}^3$, (leucocytosis). Peripheral smear showed myelocytes 7%, promyelocytes 2%, metamyelocytes 7%, blast cells 2%, band cells 8% with eosinophilia and basophilia. B HCG 1.20mIU/ml.

Emergency ultrasonography showed a heterogeneous lesion in right adnexa, 30*27mm size with ring of fire appearance, with moderate hemoperitoneum and gross splenomegaly.

A working diagnosis of hemoperitoneum with likely ruptured corpus luteal cyst was made and she was taken for emergency laparoscopy in view of hemoperitoneum.

Intraoperatively authors found 2-2.5 liters of hemoperitoneum, with active bleeding from the right ovarian tissue which was cauterized, and a biopsy was taken. Patient was transfused blood intraoperatively and postoperatively.

The histopathology report showed ovarian cyst wall lined with luteal cells. Infiltration by immature leucocytes mainly myelocytes and band cells were seen. These cells were seen within the ovarian cyst wall and also in the hemorrhagic areas (Figure 1).



Letter A represents mainly myelocytes and band cells. This is suggestive of leukemic deposits in ovarian cyst. Arrow showing ovarian cyst wall.

Figure 1: H and E staining exposes the ovarian cyst wall lined with luteal cells, infiltrate is composed of immature leucocytes.

Further IHC (immune-histochemistry) staining was done, rare CD34 positive cells (blasts) were seen within the infiltrate suggesting hematopoietic progenitor cells.

These findings were suggestive of leukemic deposits in the ovarian tissue, manifesting as a tumor of blood forming elements, there by leading to the diagnosis of extramedullary hematopoiesis of ovaries as a rare presentation of CML.

Patient was then referred to hematologist and was restarted on imatinib, with contraceptive advice and is doing well.

DISCUSSION

Chronic myelogenous leukemia (CML) is a myeloproliferative triphasic disorder characterized by the hyperplasia of fairly differentiated granulocytic cells that normally presents in middle-aged adults. Clinical manifestations of CML common to all phases of the disorder include abnormal routine blood tests, constitutional symptoms of fatigue and weight loss, abdominal pain, and abnormal bleeding episodes.

Patients with CML can rarely present with spontaneous hemoperitoneum which is commonly due to splenic hemorrhage, but hemorrhagic corpus luteal cyst can also be a cause. The differential diagnosis also includes ectopic pregnancy.

Spontaneous splenic rupture has been reported worldwide as a cause of massive hemoperitoneum in CML patients leading to acute abdomen with varying degrees of shock and even death.⁷

Corpus luteal cyst hemorrhage (though common in child bearing women), is a rare cause of spontaneous hemoperitoneum in patients of chronic myeloid leukaemia in reproductive age group. It also mimics ectopic pregnancy, along with other causes of spontaneous hemoperitoneum.⁸

Palatnik A, Narayan R, first reported a case of extramedullary hematopoiesis involving uterus, fallopian tubes, and ovaries, mimicking bilateral tubo-ovarian abscesses.⁵

Only very few cases of extramedullary hematopoiesis in genital tract have been reported till date, making it a rarer diagnosis.⁹

This case demonstrates that hematopoietic infiltration of the ovaries should also be considered in the differential diagnosis for women with CML with hemoperitonium.

This is a rare case where a hematological malignancy has presented in such a way that it needs surgical management along with medical management. This case illustrates the importance of a broad differential diagnosis when managing a patient with hemoperitoneum and the value of a multidisciplinary team approach.

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