

Case Report**Granulocytic sarcoma mimicking carcinoma of ovary, a rare case report****Abstract**

Granulocytic sarcoma in ovary is a rare haematological neoplasm due to extramedullary leukemic deposits. Here we report a case of granulocytic sarcoma of ovary in a 31 year old female with primary infertility being provisionally diagnosed as ovarian tumour. Later on, after complete haematological investigation and fine needle aspiration cytology of ovarian tumour, the case is found to be of granulocytic sarcoma associated with chronic myeloid leukemia. The aim of documenting this case report is to establish the possibility of granulocytic sarcoma in unusual site which was initially thought to be of nonhematological malignancy and proved by tissue diagnosis to be of haematological malignancy.

Key words: Chronic myeloid leukemia, extramedullary, granulocytic sarcoma.

Introduction

Granulocytic sarcoma is an extramedullary neoplasm frequently seen in acute and chronic leukaemia and myeloproliferative disorders, and is composed of primitive granulocytic cell [1]. It is a rare occurrence with an estimated incidence of 0.7 per one million in children and 2 per one million in adults [2]. It occurs in 2-8% of patients with acute and chronic myeloid leukaemia (CML) [2]. Bones, perineural tissue, lymph nodes, skin, gum and almost every site of the body is possible site for granulocytic sarcoma. But granulocytic sarcoma of ovary is a rare entity. We hereby report a case of granulocytic sarcoma of ovary in a 38-year old female of CML who was mimicking ovarian tumour due to its rarity and diagnostic dilemma.

Case report

A 31 year old female patient attended presented with lower abdominal pain and dyspepsia for 2 weeks. She had also history of found to have hypomenorrhea and primary infertility. On clinical evaluation she had pallor and splenomegaly. Clinical evaluation of other systems did not revealed any abnormality. USG abdomen and pelvis revealed splenomegaly and a right hypoechoic adnexal mass (?Multicystic enlarged ovaries). USG Doppler also showed right ovarian enlargement with predominant solid component and low resistance flow in ovarian artery, suspicious of neoplastic lesion of right ovary. Her serum CA125 was raised (56.19 units/ml). Based on imaging and biochemical parameters patient was provisionally diagnosed as carcinoma of right ovary. During routine haematological study, incidentally peripheral smear report revealed a total WBC count of 1.4 lakhs with premature cells (Myeloblasts 2% , Myelocytes 15%, Metamyelocytes 13%), suggestive of CML and diagnosis was further straightened with LAP (leukocyte alkaline phosphatase) score which was 3(normal range being 20-150). With further evaluation of ovarian status with CT scan abdomen and pelvis, patient was found to have right adnexal lesion (5×3.6cm) with splenomegaly (figure-1) and CT scan guided fine needle aspiration cytology (FNAC) of right

ovarian solid mass lesion revealed clusters of benign mesothelial cells and blood cells comprising of myelocytes, metamyelocytes, occasional promyelocyte, normoblasts, basophils and neutrophils (figure-2 and 3). Further cytogenetic analysis showed presence of Philadelphia chromosome confirming of CML. Hence this case is diagnosed as CML with granulocytic sarcoma involving ovary and started with Imatinib 400mg per day. Response evaluation after 6 months of imatinib treatment revealed complete haematological response and major molecular response. Radiologically, CT scan of abdomen and pelvis revealed disappearance of right adnexal mass with normal spleen. Now, the patient is on imatinib treatment and attended complete molecular response.



Figure1: CT scan abdomen and pelvis showing heterogeneously hypodense mass lesion in right adnexa.

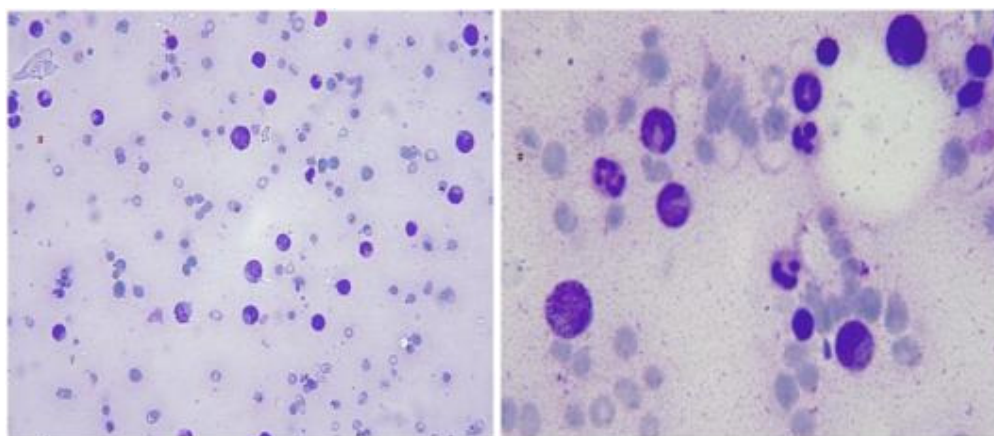


Figure2 & 3: USG guided FNAC of ovarian mass showing granulocytic precursor cells like promyelocytes, myelocytes, metamyelocytes and blast cells.

Discussion

Granulocytic sarcomas are rare extramedullary tumours consisting of premature granulocyte precursor cells. They arise de novo, or may arise in association with other hematologic disorders such as AML (acute myeloid leukemia), myelodysplastic syndrome or

myeloproliferative disorders. Granulocytic sarcoma can arise anywhere in body; it frequently involves bones, perineural tissues, lymph nodes, skin and gums. However granulocytic sarcoma in female genital tract is uncommon. Ovarian granulocytic sarcoma is a rare entity in daily practice [3]. Primary ovarian presentation of granulocytic sarcoma is very rare [4]. Granulocytic sarcoma can occur in female genital tract and may be the first clinically significant manifestation of a haematological malignancy [5]. It is further rarer to diagnose granulocytic sarcoma by FNAC [6]. This case is reported because of its uncommon location and dilemmatic way of presentation where provisional diagnosis was thought of as ovarian tumour. In this case, the aspirate obtained from ovary (CT-guided) is of paramount importance because it is showing evidence of myeloid differentiation. It is arising high index of suspicion of granulocytic sarcoma because our patient presented with infertility and lower abdominal pain as primary symptom and establishment of haematological malignancy i.e. CML. Besides, after 6 months of Imatinib treatment, CT evaluation of abdomen and pelvis was showing complete disappearance of ovarian pathology which indirectly but strongly supports the ovary being presented as granulocytic sarcoma in CML.

CML can be associated with granulocytic sarcoma. Most frequently, the tumour presents later in the natural history of CML as the disease progresses. Patients in relapse after chemotherapy or stem cell transplantation can also present with granulocytic sarcoma [7]. A granulocytic sarcoma that develops with chronic phase CML confers a worse prognosis i.e. higher risk of rapid blastic marrow transformation [7]. Granulocytic sarcoma can occur in ovary even though rare and it can mimic ovarian tumour as in this case. A granulocytic sarcoma associated with CML responds to imatinib treatment which is evident in this case in which there is disappearance of ovarian pathology after 6 month course of Imatinib treatment.

Conclusion

Granulocytic sarcomas are rare haematological neoplasms due to extramedullary leukemic deposits, and primary granulocytic sarcoma of ovary even further rare. Granulocytic sarcoma associated with CML responds well to imatinib treatment. Hence a strong vigilance is required to suspect ovarian involvement in CML as granulocytic sarcoma. Patient having CML with ovarian involvement should be evaluated to rule out the possibility of granulocytic sarcoma.

References

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