

Clinical report

Primary granulocytic sarcoma: a rare presentation of an ovarian mass

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Background: Primary granulocytic sarcoma (GS) only rarely presents with an ovarian mass classified as extramedullary myeloid progenitor tumor cells. Few patients with cases of primary GS survive after systemic chemotherapy.

Objective: To present a case of primary GS treated with complete course of chemotherapy. The patient is currently in complete remission.

Methods: Retrospective review of medical records and comparison with previous case reports.

Results: A 41-year-old woman presented with an ovarian mass of 6 months duration. There was no previous history of hematologic disease or presence of leukemic cells in other organs at the time of diagnosis. Immunohistochemistry confirmed the diagnosis. She received complete course of chemotherapy, after which complete remission was achieved.

Conclusion: A case of primary granulocytic sarcoma of the ovary was successfully treated with chemotherapy. This disease needs careful diagnosis and appropriate treatment for a good outcome.

Keywords: Chemotherapy, granulocytic sarcoma, ovarian mass

Granulocytic sarcomas (GS) are categorized as solid tumors and consist of myeloid leukemic cells. GS can present in different organs including in bones, the central nervous system, and soft tissues [1]. Lymph nodes and skin are two common sites. However, findings in genital organs are rare [2]. There have been few cases of female patients presenting with GS of the ovary. Criteria for definite diagnosis are complicated based on previous studies.

Differentiating GS from other hematologic malignancy such as lymphoma from clinical presentation and progression in patients is difficult. At present, immunohistochemistry is essential to distinguish GS from microscopic tissue morphology. Constantinou et al. [4] reported that a patient with GS of ovary did not demonstrate the outcome of treatment and prognosis of this disease. Primary GS is classified as an extremely rare condition [2, 3]. GS has been diagnosed in other genital organs, and may

result in a patient's disease-free survival [4, 5]. Nevertheless, except for a few cases, scarce data exist regarding the prognosis of patients diagnosed with primary GS with or without complete anticancer treatment.

Case report

A 41 year-old woman presented with a lower abdominal mass of 6 months duration on January 2008. Vaginal examination revealed a firm fixed mass on the right ovary. Ultrasound imaging showed a solid well-circumscribed mass in front of the uterus. The uterus and cervix were normal (**Figure 1**). The patient underwent a left salpingo-oophorectomy with omentectomy. Gross examination revealed presence of an 18 cm × 15 cm × 10 cm (1,210 g) ovarian mass. No paraaortic or iliac lymph nodes were palpated between the level of kidney and bifurcation of aorta. Liver, spleen, gall bladder, and pancreas appeared normal.

A frozen section of the ovarian mass showed a light grey-brown homogeneous solid tissue and hematoxylin and eosin stained paraffin-embedded

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sections revealed diffuse mixed small and large sized cells along with scattered immature and mature eosinophils (**Figure 2**). Multiple immunohistochemical analyses were obtained including staining positive for myeloperoxidase, CD45, CD43, but these cells tested negative with vimentin, pancytokeratin AE1/AE3, inhibin, CD 3, and CD 20 (**Figure 3**). Complete blood count was within normal limits with a normal differential count. Bone marrow aspiration and biopsy did not show presence of any myeloid leukemia. Subsequent reevaluation showed residual mass during the second month postsurgery, which indicated that the ovarian mass became progressive again (**Figure 4**).

A course of chemotherapy with cytarabine (Ara-C) and idarubicin was administered for 7 and 3 days, respectively, to reduce the ovarian mass. A month

after the first course of chemotherapy, Ara-C and mitoxantrone were given, after which patient developed febrile neutropenia (hemoculture revealed *Aeromonas veronii*). Response to treatment was successful. Chemotherapy with 2 courses of high-dose of Ara-C (HIDAC) was continued. A month after completing the second HIDAC course, work-up to evaluate GS revealed that the ovarian mass had disappeared. There was no mass or lymphadenopathy noted on both clinical examination or computed tomography (CT) of the abdomen and pelvis. Four years after chemotherapy, she continues to do well and the ovarian mass has completely resolved. No mass or any new lymphadenopathy was found on follow-up CT scan. This patient was considered as completely in remission after chemotherapy.



Figure 1. Longitudinal abdominal ultrasound images of lower abdomen. Ultrasound images of lower abdomen shows a large well-circumscribed solid mass at anterior to uterus (arrow).

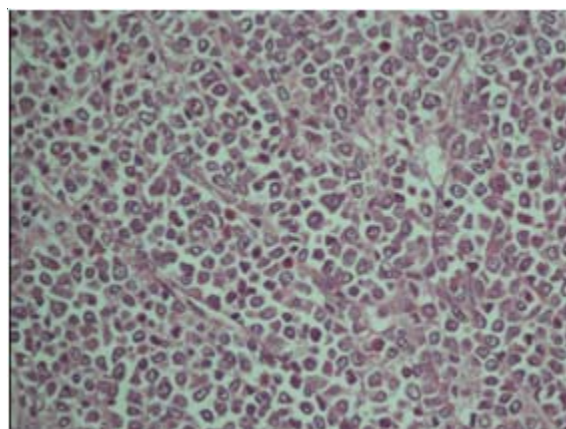


Figure 2. Microscopic examination reveals diffuse mixed small and large size oval cells, scanty cytoplasm of myeloid tumor cells. Along with scattered immature and mature eosinophils (hematoxylin and eosin staining, $\times 400$).

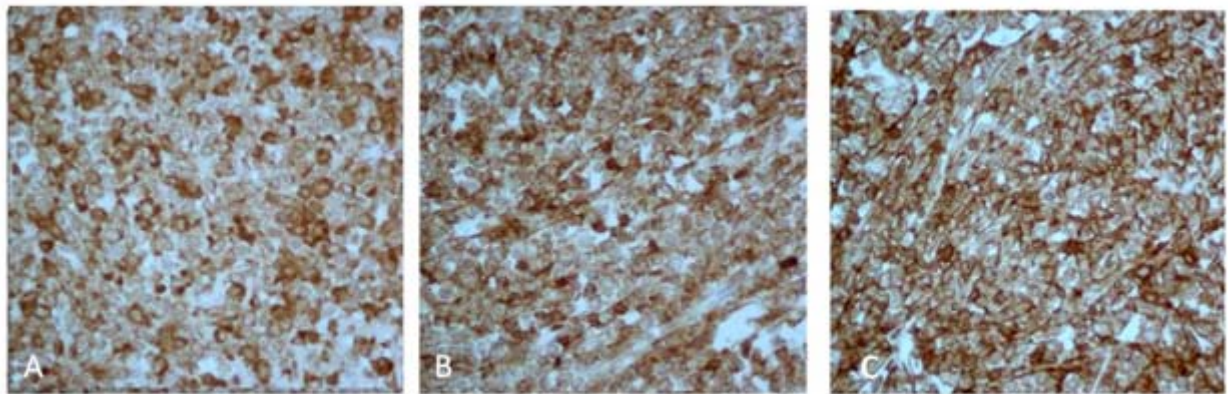


Figure 3. Tumor cells reveal intense staining for myeloperoxidase (A) ($\times 400$), CD45 (B) ($\times 400$), and CD43 (C) ($\times 400$).



Figure 4. Abdominal computed tomography. Abdominal computed tomography with intravenous contrast medium shows a recurrent ill-defined hypoattenuation mass in right lateral wall of uterus. Minimal free fluid is demonstrated in left pelvic cavity (arrows).

Discussion

We report the case of a 41-year-old woman who presented with a right ovarian mass and abdominal distension. The site of presentation and clinical response following systemic chemotherapy in GS of ovary has rarely been documented, barring a few case reports.

Extramedullary leukemia may be found in the following three conditions: (1) mass without presence of systemic leukemia, similar to our patient (2) simultaneously with blood and bone marrow with leukemic cells, and (3) as an isolated recurrence of acute myeloid leukemia or chronic myeloid leukemia [2]. Primary GS without systemic leukemia is a rare condition [1]. Primary presentation of GS in the female genital tract is extremely rare; most cases present gradually as a mass as previously reported [1-3, 6-8].

There have been only 7 reported cases of primary GS at initial diagnosis in nonleukemic patients. Cells of GS are thought to originate from intramedullary areas of the bone marrow spreading via the Haversian system throughout the entire body [5, 9]. Diagnosis is difficult and GS can be misdiagnosed as another hematologic malignancies [7]. Physicians may confuse this with lymphoma and other solid tumors. Diagnosis can be made by finding typical slightly smaller cells with finer chromatin along with myeloid differentiation. Immunohistochemical staining such as myeloperoxidase and CD45 staining are useful because they are highly specific for diagnosis. CD43 staining, most often indicate the tumor cells as a T-cell marker, which is also expressed by nearly all GS cells [7, 10, 11].

Treatment was similar to that for leukemic patients given early chemotherapy as previously reported

because patients with primary GS can develop systemic leukemia after diagnosis without any chemotherapy [7, 10, 11]. In addition, previous studies found that patients with primary GS were at high risk of developing acute leukemia within a year [2, 3]. Our patient received a chemotherapeutic regimen for acute myeloid leukemia and developed febrile neutropenia on her second course. As demonstrated by Sreejith et al. [1], Mehta et al. [2], and Jung et al. [7], primary GS may be treated by systemic chemotherapy with good response (remission ranged from 5 to 48 months). On the other hand, Pressler et al. showed that two patients underwent surgical procedures without chemotherapy and had a long complete remission (12 months) [6].

Awareness of the clinical presentation of primary GS in patients is needed for pathological diagnosis and treatment. In patients without preexisting hematologic disease, complete chemotherapy treatment similar to that for systemic leukemia is needed, along with a long period of follow-up. It will be interesting to evaluate the prognosis of primary GS in future studies.

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