CASE REPORT

Primary Granulocytic Sarcoma of Ovary

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Introduction

Granulocytic sarcomas (GS) is a rare extramedullary tumor composed of myeloid progenitor cells. Bones, lymph nodes, and skin are the favored sites of involvement. The orbit, sacrum, para-nasal sinuses, spine, and sternum are frequent sites and multiple other sites of involvement have been reported [1]. Primary involvement of the ovary is exceedingly rare leading to underdiagnosis of GS [2] and hence our interest to report this rare event.

Case Summary

A 30-year-old lady from Tamil Nadu, South India, para 2, presented with complaints of an abdominal mass and abdominal pain of 45 days' duration. She also had polymenorrhea for the preceding 2 months. General examination was normal. Per abdominal examination showed no palpable liver or spleen and there was a mass corresponding to a 20-week-sized gravid uterus arising from the pelvis. Per vaginal examination revealed a retroverted normal-sized uterus and a 10 × 10 cm firm, globular non-tender mass, felt mostly through the posterior fornix with transmitted mobility. An ultrasonography scan was done which revealed an 11 × 11 cm solid mass posterior to the uterus. The mass was very vascular with a pulsatility index of 1.0 and a resistance index of 0.5. There was negligible free fluid in the pelvis. A CT scan done elsewhere reported a large lobulated adnexal mass in the region of the rectouterine pouch, probably a broad ligament fibroid. The uterus and right ovary were reported to be normal and the left ovary was not seen separate from the mass. Serum tumor markers were normal and a staging laparotomy was planned for the patient with the diagnosis of probable malignant ovarian mass.

Intraoperatively, no free fluid was present in the peritoneal cavity. A tumor of the size 25 × 20 cm arising from the left ovary was encountered. The uterus was bulky and the uterine cavity was normal on cut section. The mass on cut section showed solid tumor tissue with necrosis and hemorrhages and did not have any specific color. The

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capsule was breached. The frozen section was reported as a malignant neoplasm, not suggestive of epithelial origin. Both tubes and the right ovary were normal. Staging laparotomy with total abdominal hysterectomy and bilateral salpingo oophorectomy and infracolic omentectomy were performed. There were no palpable lymph nodes in the pelvic or para aortic regions and the liver and spleen were normal; there were no other metastatic deposits in the abdomen or pelvis and peritoneal washings were reported to be free of cancer cells. There was a residual tumor tissue measuring 5×2 cm adherent to the lateral and posterior aspect of the right uterosacral ligament and closely adherent to the pelvic side wall, right ureter, and underlying vascular structures, which could not be resected without major injury to the ureter and vascular structures. The postoperative period was uneventful. The patient was discharged on the 7th postoperative day.

Histopathologic examination of the surgical specimen showed sheets and cords of a medium-sized relatively monomorphous population of cells with oval to irregularly contoured nuclei, fine chromatin, inconspicuous nucleoli, and scanty cytoplasm (Fig. 1). The immunohistochemical profile of the tumor cells was as follows: positive for CD43 (Fig. 2) and Myeloperoxidase and negative for alpha inhibin, CD20, and CD3. The diagnosis was *granulocytic sarcoma of ovary*.

Complete blood count and bone marrow biopsy were done which showed no evidence of primary malignancy, and thus this was considered a sporadic event in the ovary. This case was subsequently managed in conjunction with the Department of Hematology and considering the histopathology report and suboptimal cytoreduction, the patient was considered for postoperative chemotherapy. In view of localized granulocytic sarcoma, the ideal chemotherapeutic regime that was offered to her was 3 days of doxorubicin and 7 days of cytosine to be followed by high dose arabinoside and

Fig. 1 $\times 400$, H&E, sheets of medium-sized tumor cells with fine chromatin; 1–2 nucleoli and eosinophilic cytoplasm

Fig. 2 $\times 400$, CD43 immunostain, Tumor cells showing cytoplasmic positivity for CD43

cytosine (HIDAC) for 3–4 cycles. However, due to financial constraints, she was alternatively started on Hydroxy Urea 500 mg and asked to be on follow-up. On follow-up, 3 months after her surgery, she was doing fine.

Discussion

Granulocytic sarcoma is a rare solid extramedullary tumor consisting of granulocytic precursors. The tumor was originally named “chloroma” because of the green color produced by the presence of myeloperoxidase [1]. Granulocytic sarcoma is reported in 3–9 % of patients with acute or chronic myelogenous leukemia [3]. The tumor occasionally antedates the onset of systemic leukemia and occurs in a non-leukemic patient. There appears to be no sex predilection and a majority of the cases occur in the pediatric population. Although the lesion can occur at any site, bone and nervous tissue, particularly the orbit and the epidural space, are frequently involved [3–6]. An ovarian mass presenting as the initial manifestation of leukemia is extremely rare [7, 8]. Granulocytic sarcoma is not usually included in the differential diagnosis of ovarian tumors because of the rarity and poor recognition of this entity. What makes this case interesting is the fact that this was a primary sporadic event in the ovary presenting as a primary ovarian mass as the initial manifestation of this hematological tumor.

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