

Meigs' Syndrome Associated with Giant Granulosa Cell Tumor of the Ovary

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Presentation of rare occurrence of Meigs' syndrome associated with giant granulosa-cell tumor of the ovary. Giant granulosa-cell tumor of the ovary, in a patient presenting with ascites, and bilateral pleural effusion, which rapidly dried up on excision. Meigs' syndrome should always be kept in mind, not to mistakenly up-stage a patient, presenting with ascites and pleural fluid along with an ovarian mass
(*Gynecol Obstet Reprod Med* 2006; 12:000-000)

Key Words: Meigs' syndrome, Granulosa cell tumor

Sex cord-stromal tumors account for 5%-10% of all ovarian cancers; most of these (70%) are granulosa cell tumors (GCTs), which are low-grade malignancies with a relapse rate of 10%-33%. The average time to recurrence is between 5 to 10 years, although there is a case of recurrence as late as 37 years after diagnosis.¹ 10-year survival is mostly reported to be 90% for stage I and 0%-22% for stage III. The cases of bilateral ovarian involvement have an unexpectedly poor prognosis. Fox et al. in their series have reported that all seven stage Ib cases had recurred.² Meigs' syndrome, is an uncommon clinical entity characterized by ascites, hydrothorax, and an ovarian tumor that was originally believed to be specifically a fibroma; however other types of ovarian tumors such as Brenner tumors, and Krukenberg tumors are now known to be associated with this syndrome. Although the cause of Meigs' syndrome is not fully understood it is thought that hydrothorax occurs by certain lymphatics through the diaphragm. After removal of the ovarian neoplasm, there is a prompt resolution of both abdominal and pleural fluid.^{3,4}

In a review of the literature, there are only two cases^{5,6} to our knowledge, other than the herein presented case of Meigs' syndrome associated with GCT of the ovary.

Case Report

47 years old, premenopausal virgin presented to our clinic with the complaints of anorexia, hematemesis, and diarrhea of approximately 3 weeks duration. Pelvic examination revealed an adnexal mass on the right side. Computerized tomography scan and chest X-Ray Figure 1 showed an adnexal mass up to the level of the renal hilus, and bilateral pleural effusion, uterus and left ovary were normal. Thoracentesis

was negative for tumor cells. Her hemoglobin level was 6.4 gr/dl on admission and was raised to 12.4 gr/dl after serial blood transfusions. Gastroscopic, colonoscopic, and cystoscopic studies revealed no pathologic findings. Ca-125 was 553 U/ml, LDH was 502 IU/L.

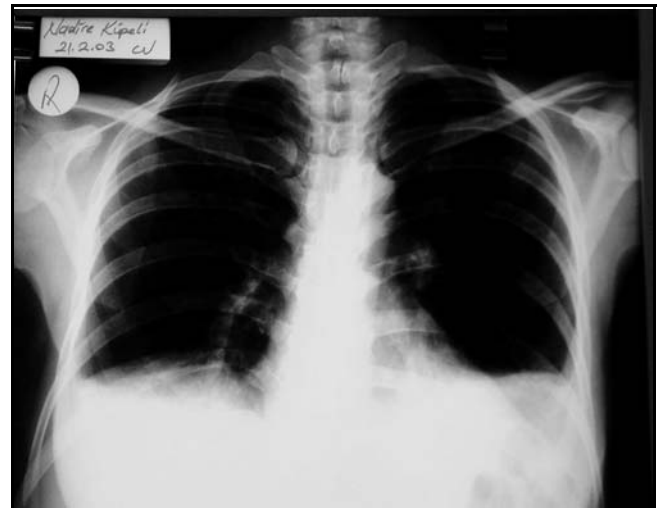


Figure 1. Chest X-Ray showing bilateral pleural effusion. Thoracentesis was negative for malignancy



Figure 2. Granulosa cell tumor of the right ovary.

Laparotomy was undertaken where a right-sided giant, partially necrotic ovarian mass, along with 150 cc of ascitic

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Submitted for Publication: 31.01.2005

Accepted for Publication: 05.02.2005

fluid admixed with blood and bilateral double ureters were noted. No signs of metastatic disease in the abdomen were found. The mass (Figure 2) was excised and sent for frozen section analysis along with ascitic fluid. Pathologic diagnosis was GCT. Total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic and paraaortic lymph node sampling, omentectomy, and appendectomy were performed. Sample of ascitic fluid was also sent for pathologic review. After pathological examination, no tumoral metastasis was found. In cross sections of the 30 X 28 X 13 cm right ovary, tumor cells showed call-exner bodies and scant cytoplasm but no mitotic figures. Hydrothorax rapidly dried up on excision, and postoperative follow-up was unremarkable. The patient, after 2 years follow-up, is still alive without disease.

Discussion

Meigs' syndrome is a rare finding in patients diagnosed with ovarian fibroma, it is even rarer associated with GCTs. In Dockerty and Masson's series this syndrome was found in only 2 of 283 patients with an ovarian fibroma.⁷ Nevertheless, it is important to keep this syndrome in mind to dismiss the possibility of a false impression that the patient has a far advanced malignancy.

GCTs occur in premenopausal women in approximately the same frequency as it occurs in postmenopausal women. Most GCTs produce estrogen although a few are androgenic; in fact they are the most common tumor that produces estrogen. The high incidence of endometrial stimulation caused by these tumors are reflected by the fact that in up to 50% of cases there is hyperplastic endometrium, and approximately 10% of patients with this tumor harbor an endometrial carcinoma, usually of the well-differentiated type. The association is twice as common in postmenopausal women. Based on these rates any postmenopausal woman with endometrial hyperplasia who is not receiving estrogen therapy, and not obese should be evaluated with the suspicion of having a granulosa-theca cell type functioning ovarian tumor.⁸

On gross examination, GCTs are usually solid and cystic, with areas of hemorrhage similar to the findings in our case. Histologically the cytoplasm is usually scanty and the typical coffee bean grooved cells are present. Although GCTs are divided into two general categories as well differentiated and moderately differentiated on the basis of their differenti-

As Cronje et al. pointed out, for the differential diagnosis of GCTs includes several conditions with an extremely poor prognosis such as poorly differentiated adenocarcinoma, and breast carcinoma metastatic to the ovary, an accurate histological diagnosis is crucial.⁹

Being the third reported case of GCT of the ovary associated with Meigs' syndrome, this unique clinical entity should be considered in the diagnosis of ovarian tumors.

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