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Misleading Diagnosis of Dysgerminoma in a Young Asymptomatic Patient

Maharjan O, Bajracharya N, Dangal G, Karki A, Pradhan HK, Shrestha R, Bhattachan K, Tiwari K, Bharati S, Maharjan S

Department of Obstetrics and Gynecology

Kathmandu Model Hospital,

Kathmandu, Nepal.

Corresponding Author

Ostha Maharjan

Department of Obstetrics and Gynecology,

Kathmandu Model Hospital,

Kathmandu, Nepal.

E-mail: osthamaharjan@gmail.com

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ABSTRACT

Dysgerminomas account for approximately one third of all malignant ovarian germ cell tumors (tumors arising from ovarian germinal elements) and are the most common ovarian malignancy detected during pregnancy. They are the only germ cell malignancy with a significant rate of bilateral ovarian involvement that is 15-20 percent. They have a variable gross appearance, but in general are solid, pink to tan to cream colored lobulated masses. They have the best prognosis of all malignant ovarian germ cell tumor variants. Two thirds are stage I at diagnosis, and prognosis is excellent even for those with advanced disease due to exquisite tumor chemosensitivity. The 5 year disease specific survival rate approximates 99 percent.

This is a case report of a huge ovarian dysgerminoma in a young unmarried lady that was quite asymptomatic. She underwent laparotomy with right ovarian cystectomy.

KEY WORDS

Dysgerminoma, Germ cell tumor, Malignant, Pvarian malignancies

INTRODUCTION

Malignant ovarian germ cell tumors (OGCT) account for less than 5% of all ovarian malignancies. Although rare, dysgerminomas are the most common, accounting for approximately 33% to 38% of all malignant ovarian germ cell tumors.[1] Dysgerminoma is composed entirely of germ cells that show morphologic and histochemical similarity to primordial germ cells. They are female analogous to male seminoma and most commonly arise in adolescents and young women, mostly in the second and third decades. Eighty-five percent of patients with dysgerminoma present with unilateral disease, and the majority of patients have stage Ia disease at the time of diagnosis.[2] While pure dysgerminomas secrete no hormones, greater than 50% have elevated lactate dehydrogenase (LDH) and placental alkaline phosphatase (ALP). Up to 5% produce beta Human Chorionic Gonadotrophin (β-hCG) due to the presence of syncytiotrophoblasts.[3] Upfront fertility-sparing surgery even with later stage disease is common due to the uniquely chemosensitive nature of OGCTs, with dysgerminomas being the most chemosensitive.[4] The chemotherapy regimen of choice is bleomycin, etoposide, and cisplatin (BEP).[5]

CASE REPORT

Miss LS is a 23 year lady presented with 3 months history of abdominal swelling. The swelling was gradual in onset, increasing in size and associated with pain and discomfort in the upper abdominal region. There was no history of nausea, vomiting, lower abdominal pain or decreased appetite. There was no associated change in bowel habits, no urinary symptoms or change in menstrual pattern. Neither there was history of shortness of breath, chest pain, weight loss, facial or limb swelling, palpable nodules in any other body parts noted.

Physical examination revealed a thin built lady. She was not pale, icteric, cyanosed or dehydrated. Her vital signs were within normal limits. Her respiratory and cardiovascular systemic findings were unremarkable. On abdominal inspection, abdomen was distended up to the level of

umbilicus. All the quadrants of abdomen moved equally with the respiration. No venous dilatations were noted. On palpation, a firm mass 26 weeks size was noted which was mobile side to side with limited top to bottom mobility. The mass was non tender, had smooth and regular margins. Pelvic examination was not done, as the patient was unmarried and sexually inactive.

Abdominal sonography revealed a large heterogenous mass measuring 20 x 18 x 17 cm in the abdominopelvic region. MDCT abdomen and pelvis (fig. 1a and 1b) revealed large well defined hypodense mass approximately measuring 24 x 16.4 x 20.3 cm noted in the abdominopelvic region extending superiorly to umbilical region. After the IV contrast, there was heterogenous enhancement of the mass with non enhancing central areas of cystic/necrotic changes. The feeding vessels were noted from the right adnexa with multiple tortuous vessels at right adnexal region. The mass was abutting the uterine fundus, urinary bladder and displacing the small bowels superolaterally. Anteriorly, the mass was abutting the anterior peritoneum and posteriorly, it was minimally compressing the inferior venacava and abdominal aorta. There was no invasion to the surrounding structures. No lymph nodes were enlarged. Minimal free fluid was noted in the pelvis. The uterus was normal in size and shape with no focal lesions. All these findings suggested the diagnosis of right adnexal cyst with differential diagnosis of ovarian fibroma, pedunculated uterine fibroid with minimal pelvic ascites.

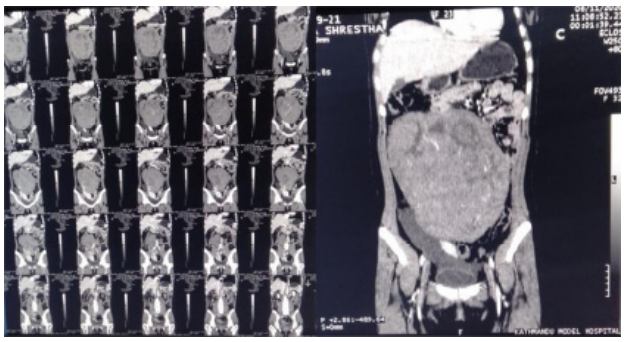

Figure 1a. MDCT abdomen-pelvis coronal plane

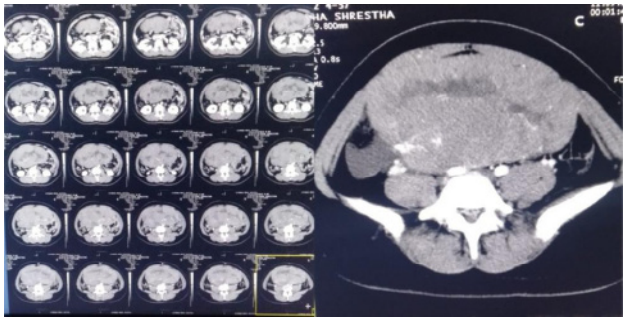

Figure 1b. MDCT abdomen-pelvis transverse cut-plane

Her basic blood reports were normal, but her tumor markers were only slightly raised [LDH- 738.0 U/L (313-618); CA 125- 134 U/ml (5-35); AFP- 1.6 ng/ml (<10); β-hCG- 34.8 (<2)]. A diagnosis of ovarian fibroma was made and she was prepared for exploratory laparotomy. She underwent laparotomy with right ovarian cystectomy. Intraoperative findings included Right ovarian tumor ~20 x 15 cm in size solid with smooth, shiny and congested external surface, no ascites, no adhesion, right fallopian tube was normal, left ovary and fallopian tube looked normal (fig. 2 and 3). Cut section of the mass showed whorls of muscle which was pale in colour. She was discharged stable and was advised to follow-up at 1 week.

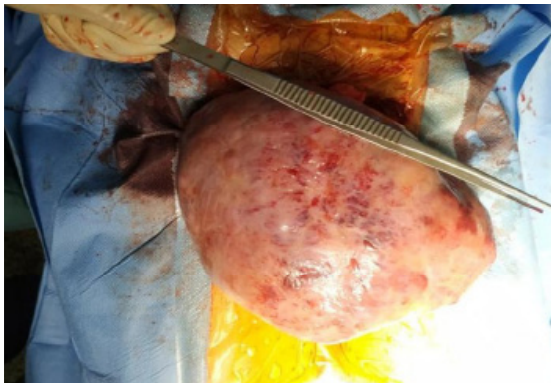

Figure 2. Comparing the tumor with 9½ inches straight Debakey Forceps

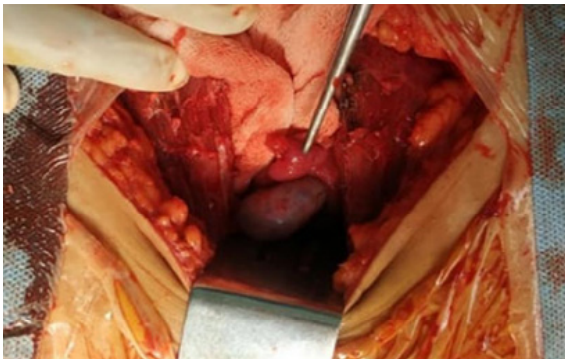

Figure 3. Normal left ovary and left fallopian tube

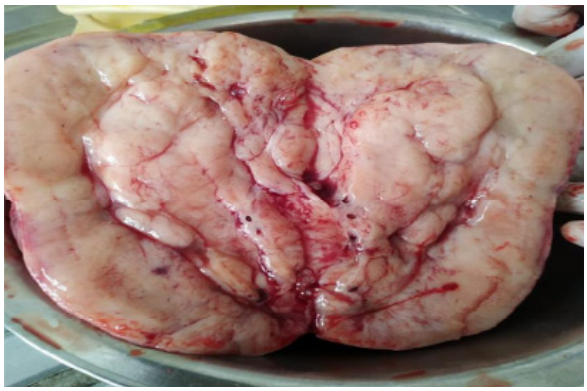

Figure 4. Cut section of the tumor

At follow-up, the histopathology reported dysgerminoma, however, the peritoneal wash was negative for malignant cells. Patient was well counseled about the disease and incompletion of the surgical management. She was advised for further staging laparotomy or chemotherapy. She opted to have second opinion at oncology center and was hence referred to Gynecologic oncologists. Telephone follow-up

was done. She received 1 cycle of adjuvant chemotherapy with BEP and has been planned for total of 3 cycles.

DISCUSSION

For several decades, radiation therapy was the traditional postoperative treatment for patients with metastatic dysgerminoma.[6,7] Although the cure rate with such treatment was excellent, irradiation usually produced ovarian failure. In 1984, treatment of the patients began with metastatic dysgerminoma with the chemotherapy combination of bleomycin, etoposide, and cisplatin (BEP) in an effort to achieve equal efficacy to that of radiotherapy while preserving fertility in these young and often nulligravid girls and women.

Killic et al. conducted a study that included 18 ovarian dysgerminoma patients with staging laparotomy concluded that fertility sparing surgery should be the choice of treatment in patients with pure ovarian dysgerminoma.[8] In addition, staging surgery, including retroperitoneal lymph node dissection is obligatory for determining stage IA patients who are exempt from adjuvant chemotherapy. Close surveillance policy enables early detection of patients with recurrences in whom salvage therapy is highly curable. In our case, due to limited surgery owing to misleading diagnosis, patient was advised for adjuvant chemotherapy.

Also in one case study, a 23 year pregnant woman was diagnosed with ovarian dysgerminoma who delivered a healthy baby and had fertility-preserving surgery, followed by 6 cycles of chemotherapy. The study showed that the long-term outcome of patients with ovarian dysgerminoma during pregnancy is excellent. Good reproductive function and high survival rate can be achieved in patients treated with conservative surgery and adjuvant chemotherapy.[9]

Ovarian dysgerminomas should be included in the differential diagnosis for a young female who presents with non-acute lower quadrant pain, palpable pelvic mass and elevated β-HCG and LDH. In our case, we only relied on radiographic report and proceeded with provisional diagnosis of ovarian fibroma. We lacked the insight of the possibility of malignancy and hence failed to perform the appropriate surgical management. The majorities of tumors are Stage IA at the time of diagnosis and can be conservatively treated with a unilateral salpingo-oophorectomy to preserve fertility. However, as soon as the diagnosis was confirmed, no delay in further adequate management was done, and patient received adjuvant chemotherapy.

This case study demonstrated the unique characteristics of this rare type of malignant ovarian germ cell tumor, including age of presentation, symptoms, elevated lab values, and radiographic characteristics.

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