International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Ara A et al. Int J Reprod Contracept Obstet Gynecol. 2022 Jul;11(7):2031-2033
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20221691

Case Report

Yolk sac tumor, a rare and challenging ovarian malignancy: case report

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Received: 29 April 2022
Accepted: 26 May 2022

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ABSTRACT

Yolk sac tumors (YST) are rare and rapidly developing neoplasm presenting in young females. They are second most common germ cell tumor after dysgerminomas. Fertility preservation is an important concern in treatment of patients of YST. We present a case of 22 years nulliparous female with rapidly evolving abdominal mass. The patient underwent fertility preserving surgery with four cycles of post operative bleomycin etoposide and paclitaxel (BEP) chemotherapy and is fairly doing well. BEP chemotherapy has successfully improved the treatment outcomes of YST patients.

Keywords: Yolk sac tumor, Germ cell tumor, Fertility preservation, BEP

INTRODUCTION

Yolk sac tumor (YST) or endodermal sinus tumor are malignant germ cell tumor usually involving gonads and presents mostly in first two decades of life. Female ovarian germ cell tumors are relatively rare 2-3% as compared to their male counterpart which is 20 times more common. YST are second most common malignant germ cell tumors after dysgerminomas and account for only 1% of all ovarian malignancy.[1] As they present in early age and have rapid malignant potential, fertility preservation is an important concern in such patients. Early diagnosis, treatment with surgery and multiagent chemotherapy are associated with high cure rates of 81.2–90.0% even in advance disease.[2]

CASE REPORT

A 22 years old nulliparous married female presented with complains of rapidly increasing abdominal swelling and weight loss for one month with pain in abdomen, fever and vomiting for 10 days duration.

On examination patient was anemic and cachexic. Abdomen was distended and a large painful mass of approximately 25×15×10 cm both solid and cystic with evidence of free fluid was present. On bimanual examination same mass of solid and cystic consistency felt and uterus could not be felt separately.

Ultrasound showed a well-defined mass of 25×16.8×95 cm arising from pelvis reaching up to epigastrium with heterogeneously upper cystic, lower solid areas, with multiple thick septation and ascetis. On contrast enhanced computed tomography (CECT), there were no lymph nodes and no distant metastasis (Figure 1).

Serum α-fetoprotein (α-FP) was remarkably raised >3000, CA-125 was 238 whereas other tumor marker were normal, haemoglobin was 8 gm% and white blood cells (WBCs) were 13500.

Patient had developed severe pain and breathing difficulties so after multidisciplinary involvement emergency staging laparotomy was done.

Intraoperative finding was a large solid cystic mass with intact capsule, densely adherent to omentum, uterus, anterior abdominal wall and pouch of Douglas. It was originating from right ovary and right tube was stretched

over it. The left ovary, tube and uterus were normal in morphological appearance. Fertility sparing surgery in form of right salpingoopherectomy was performed and due to adhesiolysis tumor capsule got ruptured. Lymphadenectomy was not done as there were no palpable lymph nodes. Biopsies from omentum and peritoneum were taken. The surgical staging of tumor was IC.

As the tumor was large and densely adherent to the periphery there was massive intraoperative blood loss. Multiple units of blood transfusions were given and patient was shifted to intensive care unit (ICU) where she was kept on ventilatory support for initial two days and after which her post operative recovery was good.

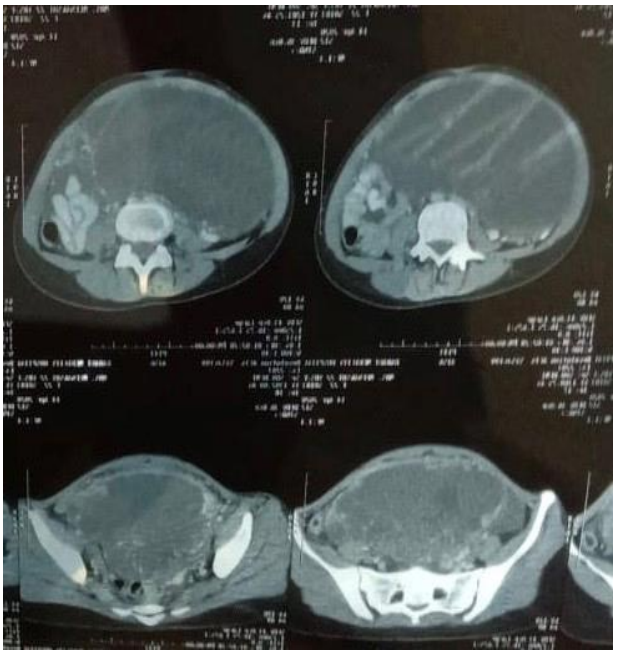

Figure 1: CECT images of large solid pelvic mass.

Histopathological finding showed tumor arranged in papillary, micro cystic and endodermal sinus pattern, lined by cells having varying amounts of clear to eosinophilic cytoplasm with nuclear pleomorphism and vesicular chromatin (Figure 2). Many Schiller Duval bodies and hyaline globules were also noted. Stroma showed myxoid and hemorrhagic areas. Omental and peritoneal biopsies were negative.

On immunohistochemistry staining there was patchy cytoplasmic expression of αFP which is gold standard in YST, other positive stainings were placental alkaline phosphatase and CK 19 (Figure 3).

The diagnosis of yolk sac tumor was confirmed and patient was referred for adjuvant chemotherapy. Presently patient has received four cycles of bleomycin, etoposide and paclitaxel (BEP) chemotherapy and is in our follow up doing fairly well. Her post chemotherapy αFP levels were 2.6.

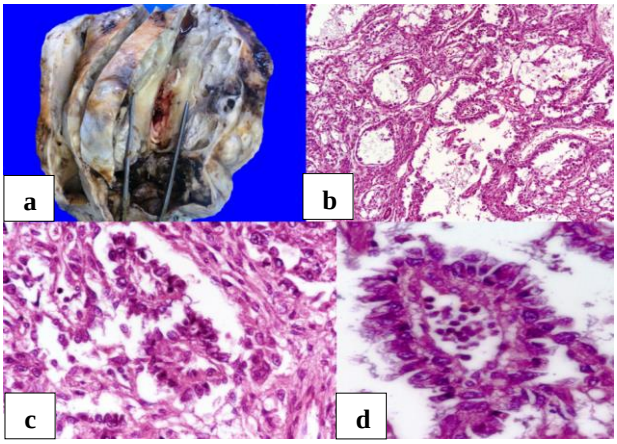

Figure 2: Gross and microscopic findings, (a) shows a large ovarian encapsulated, smooth tumour and cut surface greyish yellow to white along with areas of haemorrhage and necrosis with cystic degeneration; (b) and (c) haematoxylin and eosin-stained section shows a tumour arranged mainly in microcystic or reticular and focally in papillary pattern and lined by pleomorphic cells having varying amount of clear to eosinophilic cytoplasm and hyperchromatic nuclei. Stroma is hypocellular and having myxoid areas, and (d) demonstrating classical "Schiller-Duval bodies"; fibrovascular structure with central blood vessel surrounded by tumour cells.

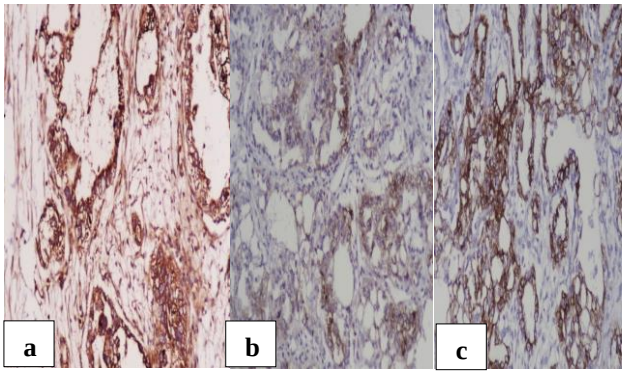

Figure 3: Immunohistochemistry staining, (a) cytoplasmic positivity for αFP, (b) focal positivity for placental alkaline phosphatase and, and (c) diffuse positivity for CK 19.

DISCUSSION

Ovarian germ cell tumor is a disease in which malignant cells arise from the germ cells of the ovary. Most of these germ cell tumors are benign and female malignant ovarian germ cell tumor (MOGCT) are rare. Since the most common age of presentations is early reproductive age group, fertility preservation is prime concern after disease removal.[1] Treating these tumors is challenging as the disease can progress rapidly, with a short tumor doubling time, and rapid spread to the peritoneum and other organs.

Most common symptoms are pelvic pain, menstrual abnormalities and abdominal mass. Sometime it may present with emergencies like torsion, hemorrhage and capsule rupture. Initial workup should include a pelvic ultrasound, tumor marker like CA-125, α-FP, serum beta human chorionic gonadotropin (S. βhCG), serum lactate dehydrogenase (S. LDH) in these patients.[2] Tumor markers specific to germ cell tumors is α-FP which is remarkably raised in these tumors. CECT is important in pre operative staging of disease.

MOGCTs are usually unilateral however; 10–15% of pure dysgerminomas are bilateral. Surgery should be unilateral oophorectomy, peritoneal washing, omental biopsy and selective removal of enlarged lymph nodes. Biopsy of a normal contra lateral ovary is not indicated. Surgery should be by an open procedure to ensure complete removal of the affected ovary with its intact tumor capsule rather than broken or ruptured.[2,3]

Operative details, histopathological findings, tumor markers and imaging studies are important for correct staging and management planning. With the advent of chemotherapy BEP, the prognosis has remarkably increased from 14% to more than 95% in advance cases of MOGCTs.[2] Patients with stage IA can be safely managed without immediate adjuvant chemotherapy. Using close surveillance, chemotherapy is reserved only for those who have relapse and patient with greater than stage IA disease, as the risk of relapse is considered high enough in such cases. The number of cycles and choice of chemotherapy regimen is based upon the histology and stage of disease. Response to treatment is monitored carefully with both tumour marker and radiological evaluation. In advance stages neoadjuvant chemotherapy should be considered followed by debulking surgery. National comprehensive cancer network (NCCN) guidelines 2017 recommends fertility sparing surgery in cases where fertility is required or complete surgical staging where fertility is not required with 3- 4 cycles of BEP chemotherapy.[4]

Post surgery tumor markers should be measured weekly until normal levels are achieved keeping in mind their half life (α-FP 5-7 days and β-HCG 48 hours). Tumor markers important for surveillance are α-FP and β-hCG along with clinical findings and radiological imaging. The chances of relapse are highest in first two years with 75% cases occurring in first year, as such intensive 4 to 8 weekly follow up is mandatory for first two years and thereafter the frequency can be decreased. NCCN recommends in YST patients with complete clinical response follow up with α-FP in every 2 to 4 months for first two years thereafter yearly for next five years.[4]

Loss of fertility is also an important concern and involvement of fertility specialist is important. Most women 87-100% regain menstrual functions and fertility within a year and 3% may experience premature menopause. Post chemotherapy negative self-image is common in young females due to hair loss, as such emotional and practical support is very important in these patients.

In a case series of 52 patients with yolk sac tumor by Rouge et al in evaluating long term fertility results, 97% patients had regular menstruation after achieving complete remission and 75% became pregnant who attempted conception only one patient suffered ovarian dysfunction.[5] In a restrospective study by Kojimara et al 83% resumed menstrual cycles and were potentially fertile and 11% had live birth rate.[2]

CONCLUSION

Yolk sac tumors are rare and aggressive malignancies mainly presenting in younger age groups. Fertility preservation is an important concern in these patients. Fertility preserving surgeries with or without adjuvant chemotherapy is the standard treatment in young patients.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Ara A, Kumari K, Rani S, Chawla I, Ahuja A, Phulware RH. Yolk sac tumor, a rare and challenging ovarian malignancy: case report. Int J Reprod Contracept Obstet Gynecol 2022;11:2031-3.