CASE REPORT

BILATERAL GIANT MUCINOUS CYSTADINOCARCINOMA OF THE OVARY: A CASE REPORT AND MANAGEMENT OPTIONS IN LIBERIA

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ABSTRACT

Background: Ovarian cancer is ranked the second most common gynecological cancer in developing countries and constitutes the fourth most common of all cancers in women. Unlike other female genital cancers, ovarian cancer has no signature symptoms or signs, and thus lacks reliable screening modalities that limit the opportunity for an early diagnosis and treatment. Case Presentation: Here, we detail the case of a 44-year-old multipara who presented with complaints of rapidly progressing abdominal swelling of 6 months' duration. The swelling was associated with abdominal pains, dyspepsia, belching, early satiety, nausea, constipation and scanty menses. An early ultrasound revealed bilateral ovarian cysts both measuring less than 2 cm. On examination, she was acutely ill looking, alert and oriented, afebrile, not pale anicteric, and had no pedal edema. The abdomen was grossly distended. There was mild generalized tenderness. Abdominal masses were not easily discernible due to marked distension and tenderness. Abdomino-pelvic ultrasound revealed a huge abdomino-pelvic multi-septated cystic mass with complex echo pattern and free intraperitoneal fluid. A CA125 assay done showed markedly elevated value (160 U/ml). An assessment of advanced ovarian tumor was made. She was prepared and planned for exploratory laparotomy surgical staging and optimum surgery. She had total abdominal hysterectomy (TAH), bilateral salpingo-oophorectomy (BSO) complete omentectomy and adjuvant chemotherapy. Conclusion: Mucinous cystadenocarcinoma of the ovary is rare among epithelial tumors. Its management is quite challenging especially in developing countries due to late presentation and lack of available diagnostic and follow up screening tools.

Keywords: Bilateral, Huge Ovarian Cystadinocarcinomas, Management, Liberia.

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INTRODUCTION

Ovarian cancer is the seventh most common cancer, and it is the most common cause of mortality from gynecological cancers globally, accounting for an estimated 239,000 new cases and 152,000 deaths annually.[1] Central and South-Eastern Europe have the highest incidence of ovarian cancer in the world (16.6 and 12.1 per 100,000,

respectively).[2] Studies have shown that African–Americans have a lower incidence of ovarian cancer, but a poorer prognosis, compared with their White counterparts. In developing countries, ovarian cancer is ranked as the second most common gynecological cancer, and constitutes the fourth most common of all cancers in women, with 17,755 incident cases in 2012.[3]

The etiology of ovarian cancer is not fully understood but commonly cited risk factors include age, family history of ovarian cancer, infertility, or use of ovulation induction agents. Factors that are protective for the disease include increasing parity, oral contraceptive use and oophorectomy. Other factors that confer weak protection for ovarian cancer include surgeries such as hysterectomy and tubal ligation, lactation and incomplete pregnancies.[-]

Unlike other female genital cancers, like cervical cancer; ovarian cancer has no signature symptoms or signs and as such, lacks reliable screening modalities, which limits the opportunities for early diagnosis and treatment initiation. As a consequence, the majority of patients with ovarian cancer present in an advanced stage, wherein the currently available treatments are ineffective.

CASE REPORT

She was a 44-year-old Liberian female, $Para_2^{+4}$, 2 alive. Her last normal menstrual period was five months prior to presentation. She presented to the outpatient department of our maternity hospital with complaints of rapidly progressing abdominal swelling for 6 months' duration. The swelling was associated with abdominal pains, dyspepsia, belching, early satiety, nausea, constipation and scanty menses. All her symptoms worsened as the abdominal swelling progressed.

Previously, at the onset of illness, the patient had presented to our hospital with complaints of moderate pelvic pain and vague abdominal symptoms. An ultrasound revealed bilateral ovarian cysts both measuring less than 2cm. She was commenced on combined oral contraceptives and antacids and advised for follow-up with serial ultrasound. However, she had been lost to follow up.

In terms of history, the patient last had childbirth 18 years ago. She had two spontaneous vaginal deliveries at term that were uncomplicated. She also had four abortions, of which three were induced and the last was spontaneously miscarried at three months, five years ago. She breastfed her children up to1 year. She attained menarche at 13 years, and her menses were regular, of three-day duration every 30-day cycle and of normal flow till the onset of her illness. She used oral contraceptives for 1 year, 20 years ago. She was treated for pelvic inflammatory diseases about ten years ago. She had no history of cigarette smoking or consumption of alcohol. She did not use talc containing powder and all her siblings are alive and well. She had no previous medical or surgical admissions, no diagnosis of chronic diseases, no known familial illness or allergies to any medication.

On examination, she was acutely ill looking, alert and oriented, afebrile, not pale anicteric, no pedal edema. Her pulse rate was 100 cycles per minute; her blood pressure was within normal range. On chest auscultation, first and second heart sounds were heard. Her respiratory rate was 20 cycles per minutes with vesicular breath sounds bilaterally. The abdomen was grossly distended. Abdominal girth was 100 cm, bulging flanks, everted umbilicus. No stigmata of liver disease [Figure 1]. There was mild generalized tenderness with demonstrable ascites

(fluid thrill). Abdominal masses were not easily discernible due to marked distension and tenderness.

Perineal examination revealed normal female external genitalia; the cervix appeared healthy. Bimanual examination was not appreciable due to huge abdominal distension. Abdomino-pelvic ultrasound revealed a huge (>25 cm x 30 cm) multi-septated cystic mass with complex echo patternand free intraperitoneal fluid. Liver, kidneys and uterus appeared normal. A Chest X Ray (CXR) also showed no features of lung metastasis. A CA125 assay done showed markedly elevated value (160U/ml), normal range 0-35U/ml. Other supportive investigations done were Quantitative Beta human chorionic gonadotropin (hCG), lactate dehydrogenase (LDH), liver function test (LFT) and kidney function test (KFT), which were all within normal limits. Hepatitis B surface antigen was also negative; haemoglobin was 9.2 g/dl, platelets was 738,000/µL (thrombocytosis).

An assessment of advanced ovarian tumor was made. She was counseled about the diagnosis and the management options. A consent for the surgery and other management options was sought and obtained from the patient. She was prepared and planned for exploratory laparotomy surgical staging and optimum surgery.

The patient was given general anesthesia with left lateral shift. A midline incision was administered after proper cleaning and draping. Bilateral huge cystic masses were noticed arising from both left and right ovaries. They consisted of cystic and solid areas, with papillae and areas of necrosis and hemorrhage [Figure 2]. There were mucoid secretions all over the abdominal cavity. The left cyst measured 40 x 30 x 25 cm. The right cyst measured about 35 x 25 x 20 cm. Their combine weight was more than 35kg. There were no clusters of cell implant on the visceral and parietal peritoneal surfaces and on the omentum. The appendix appeared normal. The hepatic surfaces were felt nodular but there were no nodular masses more than 1 cm size in both the pelvis and the abdomen. The cystic masses were clamped at their pedicles, excised and transfixed. A total abdominal hysterectomy with bilateral salpingo-oophorectomy and omentectomy was then followed. Further peritoneal exploration was ensued and copious lavage with normal saline was followed. Post-operative recovery was uncomplicated and the patient was discharged after a week. She was on regular follow up and had currently received the first course of chemotherapy three weeks after discharge. She was planned to have 6 courses of paclitaxel-carboplatin doublets given at 3-4 weekly intervals. Histology report revealed mucinous cystadenocarcinoma of right and left ovary with capsular breech. Omentum showed metastatic deposits.

Figure 1. Grossly Distended Abdomen due to Ovarian Malignancy

DISCUSSION

Based on the anatomic structures of the ovary, ovarian tumors are classified into three major categories. These are epithelial stromal tumors, sex cord–stromal tumors, and germ cell tumors. Epithelial-stromal tumors, which are the most common type of ovarian cancers, account for about 60% of all ovarian tumors and 90% of malignant ovarian tumors. Epithelial tumors are

Figure 2: Two Hugely Cystic, Multi Loculated Mucin Secreting Ovarian Masses

further subdivided into five types comprising high-grade serous carcinoma (HGSC), 70%; endometrioid carcinoma, 10%; clear cell carcinoma, 10%; mucinous carcinoma, 3%; and low-grade serous carcinoma, < 5%.[9] This patient was a 44 year old multipara and she is within the age range of presentation (33 to 83 years) as reported by Massad and colleagues.[10] We could not identify a reliable risk factor for ovarian cancer in this patient apart from pelvic inflammatory disease; however, some of the protective factors for ovarian cancers such as the use of combined oral contraceptive pills, breastfeeding, pregnancy at an early age were identified in this patient.

Mucinous cancers of the ovary are bilateral in 15% of cases; their tumor markers are carcino embryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9)[13] in most cases. This patient presented with bilateral tumor, a tumor marker, CA125 the only tumor marker that could be assessed at the facility was found to be markedly raised. This is not surprising since other workers reported raised CA125 in mucinous cystadinocarcinoma. Apart from abdomino-pelvic ultrasound scan of which she had, other imaging modalities such as CT Scan and MRI are recommended especially where there is no pelvic mass in the presence of ascites, and liver or pancreatic tumors are suspected.

Treatment of ovarian cancer is based on findings following surgical staging. In advanced ovarian cancer (stage III and IV) as in this patient, treatment consists of total abdominal hysterectomy (TAH), bilateral salpingo-oophorectomy (BSO) complete omentectomy, removal of any tumor that can be seen or palpated and resection of any metastatic lesions from the peritoneal surfaces or from the intestines with a target of not leave behind any tumor of more than 1 cm in its largest diameter, followed by immediate chemotherapy.

The mucinous tumors are filled with a mucus-like material, of which their name was driven based on this property. This mucus is produced by mucus-secreting goblet cells very similar to the cells lining normal intestine. They may become large as in this patient. The cystadenocarcinomas contain a more solid growth pattern with the hallmarks of malignancy: Cellular atypia and stratification, loss of the normal architecture of the tissue, and necrosis as seen in this patient. The mucinous ovarian cancer sometimes associated with pseudomyxoma peritonei, where the tumor associated with extensive mucinous ascites and adhesions, cluster of cell implants on the visceral and parietal peritoneal surfaces, and production of thick gelatinous mucin.[17] In this patents there were no cluster of cell implants on the peritoneal surfaces as such we favored the diagnosis of mucinous cystadenocarcinoma even before the histological diagnosis. Frozen section would have helped us with quick intra-operative diagnosis and the need for appendectomy in the case of pseudomyxoma peritonei but it was not available in the center. In this case the tumors were removed, followed by TAH, BSO and omentectomy. The lymph nodes

were not dissected since the tumor was advanced evidenced by hepatic parenchymal metastasis of which would not change the prognosis following lymph node dissection.

Standard first line chemotherapy of epithelial ovarian cancer consists of a combination of platinum based and taxane drugs. In this case we already commenced this patient on 6 courses of paclitaxel-carboplatin doublets given at 3-4 weekly intervals.

Clinical pathological prognostic factors for epithelial ovarian cancers include stage of the disease, histological grade, age of patient, cell type, status of the capsule, cellular / tumor marker prognostic factors, malignant cytology of the ascetic fluid, lymph node involvement and the amount of residual tumor after surgery. In this patient the prognosis is poor because she presented in advanced stage of the disease.

CONCLUSION

Mucinous cystadenocarcinoma of the ovary is rare among epithelial tumors. Its management is quite challenging especially in developing countries due to late presentation and lack of available diagnostic and follow up screening tools. Clinicians should have high index of suspicion despite our poor resources to avoid treatment at advanced stage of the disease.

Conflict of interest: None

Authors' contribution: AR and DK carried out the clinical assessment, and patient evaluation and subsequent management. AR drafted the manuscript; critically revised the manuscript for intellectual content. All authors read and approved the final manuscript. AR and DK are guarantors of the paper.

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