INTRODUCTION

Amongst epithelial ovarian tumors, 90% are malignant or have malignant potential. Ten percent of epithelial ovarian tumors are categorized as borderline ovarian tumors, out of these 70% are serous type and 30% are mucinous type. Borderline malignancy is difficult to diagnose on clinical examination and may cause difficulty on histopathological examination as well, particularly of the mucinous type. The hallmark of borderline ovarian tumor is non-invasion of basement membrane. Aggressive borderline tumor, is histologically characterized by proliferative epithelial buds and marked aneuploidy. Very rarely it is seen that borderline ovarian tumor may become aggressive, therefore, it is important to have definitive protocols for management of borderline ovarian tumors.

This report describes one such case with management.

CASE REPORT

A 59 years old menopausal lady presented as a diagnosed case of mucinous tumor of ovary with gross ascites. She had received chemotherapy but no response was found after third line of chemotherapy over a period of 12 months.

She was first diagnosed in February, 2010 by ultrasound that showed a well-defined multilocular cystic mass which extended into abdomen measuring approximately 13 x 8 cm with thick septations and gross ascites. These findings were confirmed on CT scan. Tumor markers levels were raised; CA125 was 66.9 IU/ml, CEA was 7.46 ng/ml; βHCG was < 1.2 mlU/ml, Alpha Fetoprotein level was 2.18 ng/ml and all other investigations were within normal limits. CT guided trucut biopsy and histopathology showed mucinous neoplasm. Ascitic fluid cytology in March, 2010 showed highly atypical mucinous cells considered malignant.

She was started on 1st line chemotherapy and was given 4 cycles of carboplatin and paclitaxel. She underwent paracentesis multiple times during chemotherapy, due to gross ascites but the fluid reaccumulated within next 2 - 3 days. Thereafter, 2nd line chemotherapy was started; 2 cycles of Injection Hycamtin (Topotecan) were given without response. Third combination chemotherapy comprising of inj. avastin (bevacizumab) and taxol (paclitaxel) was given in September 2010.

CT scan was repeated in October, 2010, but there was no change in the mass and ascites. CT scan also showed bilateral renal parenchymal disease and atrophic kidneys along with left inguinal hernia. She underwent herniorrhaphy, while her clinical situation regarding the ovarian tumor and ascites was the same. Further chemotherapy was withheld due to complications of gross ascites and cytotoxic drugs and debulking surgery was advised.

She reported to the hospital with grossly distended abdomen due to gross ascites and large ovarian tumor. Her Hb was 9 g/dl, urea was 68 mg/dl and creatinine was 1.6 mg/dl, rest of the investigations were within normal limits. Echocardiography showed signs of cardiomyopathy.

A team of medical specialist, surgeon, anesthetist, gynaecologist and the administrator was formed to evaluate the patient’s condition and disease prognosis. Family counselling was carried out. After preparing and
stabilizing the general condition and getting informed consent, exploratory laparotomy was carried out in December, 2010. Midline para umbilical approach was used. Seven liters of ascitic fluid was drained. Large left ovarian multilocular tumor without any external sub-serosal attachments was found. It was extending from epigastrium to hypogastrium containing 2 litres of mucinous material in it, which was drained. After reducing the size, tumor was pulled out and removed (Figure 1). Uterus, right ovary, liver, omentum and intestines were apparently normal including appendix. There were no peritoneal deposits and lymph nodes were not palpable. Total abdominal hysterectomy and bilateral Salpingo-Oophorectomy was performed. Her postoperative period remained uneventful and was discharged after 8 days.

Histopathology showed borderline mucinous tumor of ovary, and required no further treatment. She kept regular follow-up at 3 monthly interval and is still on 3 monthly check ups. There is no recurrence of disease, her abdominopelvic ultrasound, CT scan and CA125 performed are reported clear up till today.

DISCUSSION

It is estimated that one in every 55 females i.e. (1.8%) may have some form of ovarian cancer in her life time. Approximately 90% of these cancers are tumors of epithelial origin. Borderline ovarian tumors comprise approximately 15% of all epithelial ovarian tumors,1 are histologically defined by the presence of a complex architecture in addition to nuclear atypia and mitotic activity but most importantly, lack invasion of the underlying stroma, which distinguishes low malignant potential tumors from ovarian carcinoma. Ovarian tumors of Low Malignant Potential (LMP), or borderline ovarian cancer, was mistaken for true epithelial ovarian cancer for many decades. However, it was realized that some women diagnosed with ovarian cancer had an indolent course and their disease was different from women with invasive epithelial ovarian cancer.

The etiology of this disease remains unclear because of the small number of cases and the lack of randomized controlled studies. Based on molecular studies, some mucinous borderline tumors of the ovary may actually represent metastasis from the appendix.1 The mean age of occurrence is approximately 10 years younger than that of women with frankly malignant ovarian cancer. The most common presenting symptoms were abdominal pain, increasing girth or abdominal distension, and abdominal mass. Approximately 23% of patients were asymptomatic.

Ultrasonographic signs on transvaginal ultrasound specific to borderline ovarian tumors are, unilocular-cyst with a positive ovarian crescent sign and extensive papillary projections arising from the inner wall, or a cyst with a well-defined multicellular nodule.2 Computed Tomography (CT) scanning should be considered pre-operatively to identify possible foci of metastasis. CT scanning can also be useful when following the patient in the future.3 MRI, on account of its superior soft tissue distinction and multiplanar capabilities, allows better characterization of complex cystic masses particularly with regard to the depiction of septations and mural nodules. Mucinous borderline ovarian tumors commonly appear as multilocular cystic masses with numerous septa and contain fluids of different signal intensities on T1- or T2-weighted MR images.4

CA-125 levels are not shown to aid in the diagnosis or follow-up care of patients with borderline tumors. The Cancer Antigen-125 (CA-125) levels were normal in 65% of the recurring cases.

When a complex ovarian mass is discovered, surgery is often, if not always, indicated. Contraindications to surgery include medical reasons (i.e., the patient is too great a surgical risk secondary to other medical problems) or patient’s refusal. Otherwise, the masses should be surgically removed.

Extensive tissue sampling of the tumors is one of the most important steps involved in the process of reaching an accurate diagnosis as mentioned above, followed by a meticulous microscopic examination. One section for every 1 or 2 cm of the tumor diameter should be taken. The sections should be obtained from solid and papillary areas as well as from areas of nodular thickening, necrosis or focal hemorrhage.

Complete surgical staging is of great importance. Proper staging is defined as an exploration of the entire abdominal cavity through mid-line incision, with peritoneal washings, infracolicomentectomy, and multiple peritoneal biopsies.4 Borderline ovarian cancer
is staged according to the FIGO classification of ovarian cancer.

Total abdominal hysterectomy with bilateral Salpingo-Oophorectomy is the main step of treatment, however, fertility sparing surgery can be offered to young women.5,6 Borderline tumors have an excellent overall prognosis,6 with 60% chance of having stage-I disease. Although borderline ovarian tumors have an excellent prognosis, they are not exempt from a risk of recurrence. Patients with stage-I disease confirmed by comprehensive staging have a recurrence rate of approximately 15%. Five-year survival rate for such patients is 100%, whereas 10-year survival rate is 90 - 95%.

Long-term follow-up is required since the tumor can recur up to 20 years after initial diagnosis.7 Follow-up is usually a combination of clinical examination, transvaginal ultrasound, and CA-125 levels. During the initial 2 years follow-up evaluation is performed every 3 months. Patients are then evaluated biannually for 3 - 5 years after surgery and then annually thereafter.4

The pathologist has a pivotal role in assessment of the borderline nature of ovarian tumors and in identification of high-risk criteria, most of which are histological like intraepithelial carcinoma and presence of invasive implants.8

REFERENCES