Case Report

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Carcinosarcoma of ovary - a rare case report and literature overview

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ABSTRACT

Ovarian carcinosarcoma is one of the lethal malignant tumours of the female genital system. It is extremely rare and represents 1% of all malignant ovarian tumours. It occurs most commonly in postmenopausal women with the incidence peak in sixth decade. Our reported case was a 41 years nulliparous lady who presented with abdominal distension for one month. Physical examination revealed a huge non tender abdomino-pelvic mass suspected to be of ovarian origin. She was investigated and radiological studies confirmed a left ovarian cystic mass of 26 cm x 22 cms with solid enhancement. Calculated RMI was 66. Staging laparotomy was performed and tense smooth surface cystic mass arising from left ovary was found and removed intact. Total abdominal hysterectomy, salpingo-oophorectomy, infracolic omentectomy was done along with the peritoneal, lymph node biopsies and peritoneal washings for cytology. The presentation and findings were indistinguishable from epithelial ovarian neoplasm. Histo-pathology revealed carcinosarcoma of ovary with positive peritoneal cytology. Staged as IC according to FIGO criteria. The patient was given 6 cycles of adjuvant chemotherapy with carboplatin and ifosfamide and was in good health at the time of follow up visits.

Keywords: Ovarian carcinosarcoma, Staging laparotomy, Adjuvant chemotherapy

INTRODUCTION

Ovarian carcinosarcomas also known as malignant mixed mullerian tumour or malignant mesenchymal tumour (MMMT) are rare malignant neoplasms of the ovary accounting for less than 1% ovarian neoplasms with fewer than 400 cases in the literature. Most affected women are postmenopausal. They present with vague symptoms and are in advanced stage with wide metastasis at the time of surgery. It is a biologically aggressive tumour with poor prognosis. We report a case of carcinosarcoma of ovary stage Ic which was successfully treated with surgery and chemotherapy.

CASE REPORT

A female patient of 41 years, nulliparous with regular menstrual cycles presented with abdominal distension and heaviness of the abdomen for one month, indigestion and difficulty in breathing during lying down posture for 15 days. Past medical and family history were not contributory. General physical examination was normal. Abdominal examination revealed a tense non tender cystic mass with smooth surface of nearly 25 x 20 cms. Occupying whole of the abdomen. On pelvic examination, the lower border of the mass could be felt in the posterior and lateral fornices. Uterine size could not be made out separately. Rectal examination was normal.

Base line investigations were found normal. Ultrasound examination revealed a huge abdomino-pelvic cystic lesion of 27 x 22 cms with multiple septations, internal echoes and solid areas of size 6 x 5 cms probably arising from left ovary. MRI abdomen and pelvis confirmed the diagnosis of left ovarian multiloculated cyst with solid areas. Right ovary could not be delineated separately. Uterus appears normal. No free fluid in the peritoneal cavity and lymphadenopathy. Tumour markers, serum

CA-125 (22.2 U/ml) and CA 19-9 (13 U/ml) were within normal range. Calculated RMI was 66.6.



Figure 1: Gross specimen of left ovarian cyst.



Figure 2: Cut section of the specimen showing solid areas and 4 litres of haemorrhagic fluid.

Diagnosed as left ovarian malignant neoplasm. Staging laparotomy was performed. A tense smooth surfaced cystic mass arising from the left ovary of 28 x 22 cms was found and excised intact (Figure 1). Uterus, right ovary with fallopian tubes, omentum and all abdominal organs were unremarkable. No ascites lymphadenopathy. Patient underwent total abdominal hysterectomy, salpingo-oophorectomy, infra colic omentectomy, biopsies from para colic gutters, bladder and bowel serosa, lymph node biopsy and peritoneal fluid cytology. Gross pathology of the left ovary revealed a multiloculated cyst with thick cyst wall, haemorrhagic gelatinous material with solid and necrotic areas (Figure 2). Histo-pathology reported as poorly differentiated malignant tumour with epithelial and stromal components suggestive of carcino-sarcoma of ovary with positive peritoneal fluid cytology for malignant cells, no evidence of metastatic deposits in right ovary, uterus and omentum. Staged as IC ovarian MMMT according to FIGO criteria. Post-operative period was uneventful. Referred to medical oncologist and adjuvant chemotherapy started with paclitaxel 175 mg/m² and carboplatin AUC 5 every 3 weeks for 6 courses. She was followed up every 3 months during first year and every 4 months during second year. Pelvic examination and CA

125 were done during every visit and she is doing well till now.

DISCUSSION

Carcinosarcomas of the female genital tract also known as malignant mixed mullerian tumour occurs in uterus, cervix, vagina, ovaries and fallopian tubes in order of decreasing frequency. Ovarian carcinosarcoma accounts for less than 1% of all ovarian neoplasms. Most of them are in older age group. The presentation of the tumour is indistinguishable from epithelial ovarian cancer. Early diagnosis is very difficult because of the rarity and insidious onset. Histologically it is biphasic with carcinomatous (epithelial) and sarcomatous (stromal) elements. Carcinomatous portion predominates in the metastatic sites and thus determines the clinical course of advanced carcinosarcoma.

Our case presented at an young age with huge smooth walled multicystic mass having solid components mimicking early stage epithelial ovarian carcinoma. Histopathology revealed homogenous ovarian carcinosarcoma. The survival for both early and late stage carcinosarcoma is inferior to serous tumors of the ovary. A retrospective analytical study conducted on the long term survival of 37 cases of ovarian carcinosarcoma (30% of patients -FIGO Stage I and II) have found 40% 1-year survival and 6% 5-year survival rate.

The main stay of management for carcinosarcoma remains cytoreductive surgery followed by chemotherapy. Though there is no established consensus guideline on the optimal adjuvant chemotherapeutic regimen, review of literature supports the use of platinum containing regimens with 68% overall response rate in the platinum group compared with 23% response rate in the non-platinum containing regimens. ^{6,7} Our case received 6 cycles of adjuvant chemotherapy with cisplatin and ifosfamide.

There are limited reports in the literature to date that define the prognostic factors. Study conducted by the Gynecologic Oncology Group have found that the predictive factors for better survival were early diagnosis, presence of unilateral ovarian tumour, no pelvic lymph node metastasis, metastatic deposits less than 2cms, omentectomy, no gross residual implants after surgery, platinum treatment, cisplatin and ifosfamide regimen. Age, parity, menopausal status, primary tumour size, para aortic lymph node metastasis, pretreatment CA-125, paclitaxel was not predictive of the overall survival. Bilateral ovarian tumours and metastatic tumours larger than 2cms were significantly associated with a poorer overall survival.⁸ Regarding our case, the factors which contributed for the good prognosis were early stage disease, unilateral ovarian tumour, no pelvic lymph node metastasis, omentectomy, no residual implants after surgery and platinum based treatment.

CONCLUSION

Ovarian cancers are difficult to diagnose in early stage as most of them are either asymptomatic or present with vague generalized symptoms. Clinical and radiological findings of ovarian cancer are a major concern about diagnostic challenges. Histopathology, playing important role in such cases, would be helpful for better management of patient. Our case of carcinosarcoma was a rarity to be staged to IC and was aggressively treated with surgery and chemotherapy. This allowed the patient to remain free of recurrence.

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