Case Report

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Carcinosarcoma: A Rare Tumour of the Ovary

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Abstract

Introduction: 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, with only 10% incidence in younger women. They are associated with an aggressive clinical course and overall poor prognosis. *Case report:* Our reported case was a 60 year old postmenopausal lady who presented to us with pain and mass in lower abdomen since 15 days. Physical examination revealed a huge non tender suprapubic mass of uterine size 24 weeks suspected to be of ovarian origin. Shewas investigated and radiological studies confirmed a lesion of approx. 13.6 x 14.3 x 15.3 cms arising probably arising from the ovary, with heterogenous enhancement. Staging laparotomy was done with maximum debulking of large ovarian mass followed by Total Abdominal Hysterectomy with bilateral salpingo-oophorectomy with infra colic omentectomy. Left ovary was totally replaced by growth. Right ovary was small atrophic. Histopathology revealed Malignant Mixed Mullerian Tumour (MMMT) of the ovary with positive peritoneal cytology. *Conclusion:* The poor prognosis associated with this rare disease emphasizes the need for collaborative prospective studies targeted to better understand the molecular changes of MMMT and the need to design new therapeutic regimens to improve patients' survival.

Keywords: Malignant mixed Mullerian tumour; ovary; carcinosarcoma; staging laparotomy.

Introduction

Primary ovarian carcinosarcoma is a rare tumour. It accounts for 1-3% of ovarian malignancies. By definition, in this tumour, both epithelial and stromal components are malignant. It is also known as malignant mixed mesodermal tumouror malignant mixed Mullerian tumour. They are furthersubclassified as "heterologous" or "homologous." This categorization is based on the presence or absence of astromal component containing mesenchymal tissue notnormally found at the primary tumour site [1]. Usually, thereis the extra-ovarian intra-abdominal spread at the time of diagnosis in the majority of the cases. The primary treatment has traditionally been surgical cytoreduction, followed byradiotherapy and chemotherapy or chemotherapy alone [2]. These tumors are aggressive in nature with poor prognosis [1].

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Case Report

A 60 year old postmenopausal female presented with pain and distension of abdomen since 15 days. She came to us with complaints of white discharge per vaginum on and off, breathlessness and increased frequency of micturition and loss of appetite since 10 days. She had no history of bleeding per vaginum, loss of weight. There was no past history of any medical or surgical illness.

On examination, she was averagely built with pallor present. Her pulse, blood pressure, respiratory and cardiovascular system were normal. Her abdomen was distended. She had a suprapubic mass of 24 weeks uterine size, cystic in nature, non tender. Per speculum examination revealed cervix flushed with vagina and vagina was pale. Cervix was flushed with vagina as revealed by per vaginal examination. Uterine size could not be made out as there was a tense cystic mass felt through all fornices. Per rectal examination revealed a diffuse cystic mass felt anteriorly and the rectal mucosa was free.

The contrast enhanced computerised tomography (CT) of the abdomen showed a well defined hypodense lesion of approximately 13.6 x 14.3 x 15.3 cm noted in the lower abdomen, arising probably from the ovary. Both the ovaries not visualised separately. Uterus appears atrophic. Multiple heterogenous enhancing lymph nodes seen in the pre and para aortic region and pelvis, largest of size 1.2 x 1.2 cm. The ascites was moderate. The cytological examination of the peritoneal fluid was positive for malignant cells.

The level of cancer antigen (CA)-125 was elevated (281.3 IU/ml, normal <35 IU/ml). The routine haematological and biochemical investigations were within the normal limits. A staging laparotomy was done. Peritoneal fluid was haemorrhagic. Peritoneum was thin, vascular and adherent to the bladder on the lower half. Intra operatively there was a large irregular friable mass present just below the peritoneum, occupying the space below the umbilicus, adherent to the small and large intestine. The uterus was small and atrophic, both adnexa was oedematous and vascular. Left ovary was totally replaced by growth. Right ovary was atrophic. Maximum debulking surgery was done followed by total abdominal hysterectomy with bilateral salpingo-oophorectomy, along with Infra colic omentectomy. There were 5-6 pre and para-aortic, non-mobile, enlarged lymph nodes. Peritoneal biopsies were taken. All specimens and peritoneal fluid sent for histopathological and cytological examination.

Cut section of the malignant ovarian mass showed areas of necrosis, cystic areas, whitish nodules brownish areas with necrosis. Microscopically, there were solid sheets of plump spindle cells with ovoid hyperchromatic nuclei and high mitotic activity. There is moderate pleomorphism and anisonucleosis with many foci of poorly differentiated cells. Ocassional bizzare and multinucleolated cells are also seen. Few fragments show islands of chondroid differentiation, multinucleate and rhabdoid differentiation. Occasional focus shows squamous differentiation. At places plump spindles cells arranged in interlacing fasciles and whorl like pattern are seen. Extensive areas of haemorrhage and necrosis are seen. Atypical mitotic figures are plenty. Cervix showed chronic cervicitis with extensive squamous metaplasia and mild dysplasia. There was no evidence of metastatic deposits. Based on the histopathological findings, the final diagnosis was given as Malignant Mixed Mullerian Tumour (Carcinosarcoma) of the ovary.

The patient had an uneventful post operative period. She was discharged and advised medical oncology consultation following surgery. However, in our case, patient failed to follow up for further management.

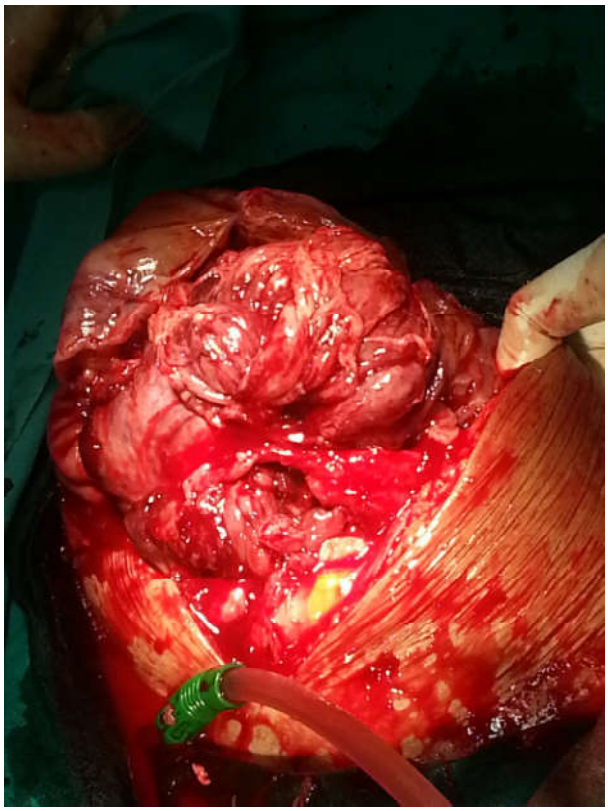
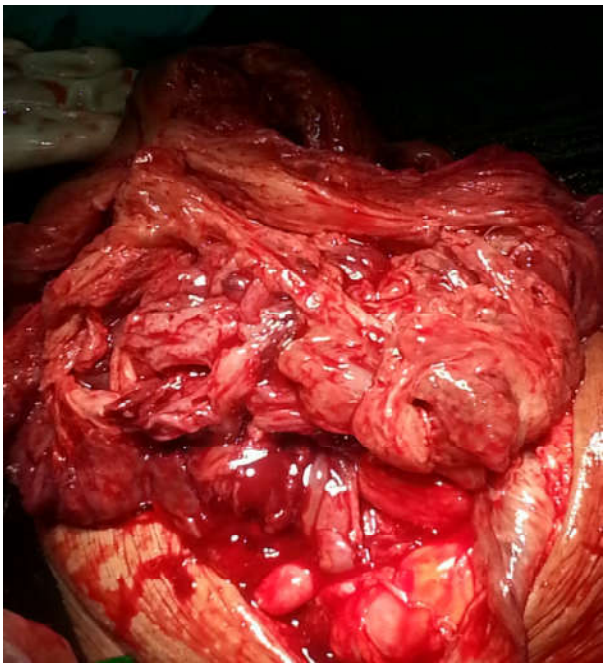


Fig. 1:



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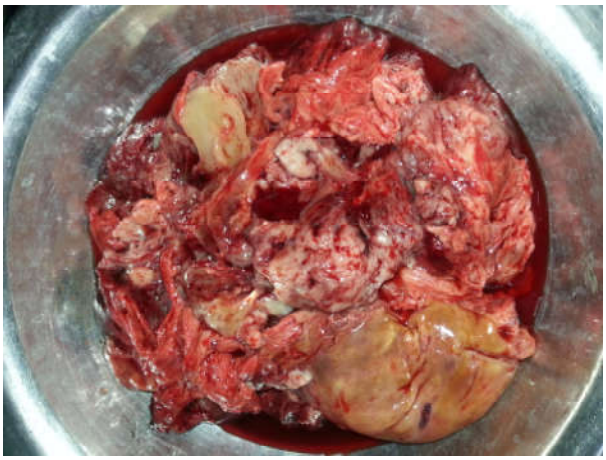

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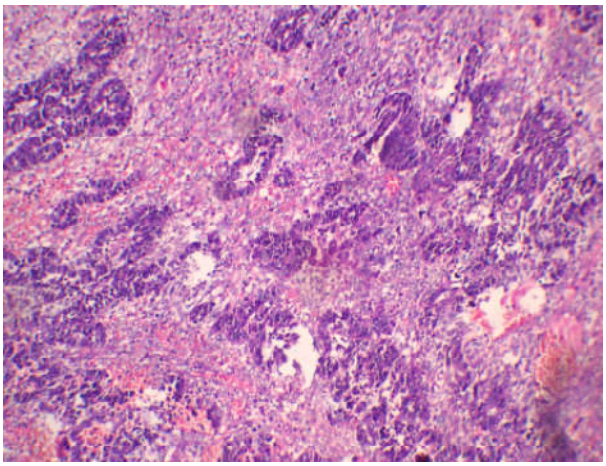

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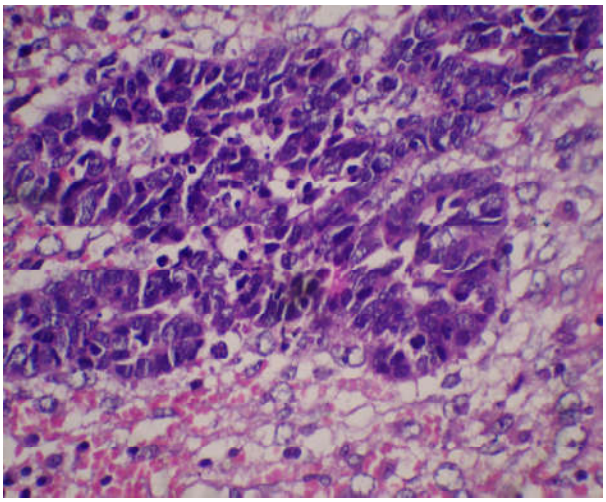

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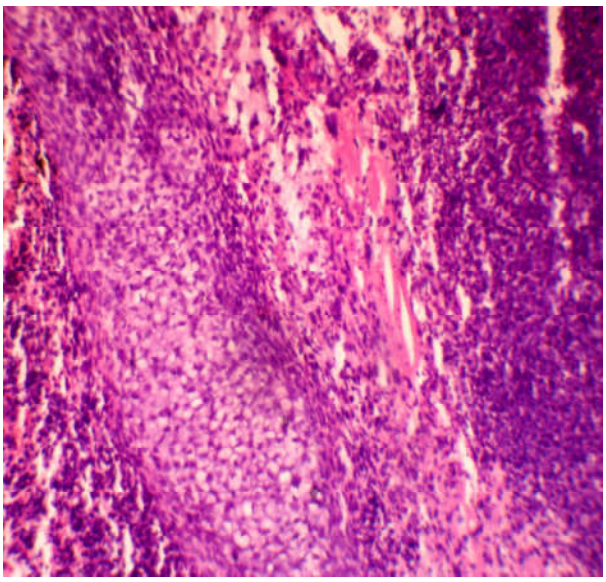

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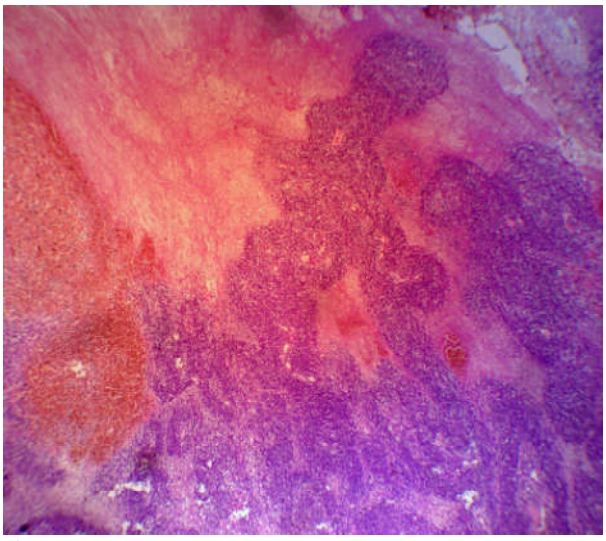

Fig. 7:

Discussion

Carcinosarcoma is an aggressive tumour, which associates some epithelial elements (carcinoma) with a stromal component (sarcoma). This tumour can be found in the female genital tract, mostly in the uterus, and it can be found even onthe ovaries but it's very rare. Malignant Mixed Mesodermal Tumours of the ovary arerare neoplasms associated with an aggressive clinical course and overall poor prognosis. The results of previous studies have demonstrated that the majority of patients with MMMT of the ovary are Caucasian and their cases are very advanced at the time of surgery [3].

Ovarian MMMT is composed histologically of malignant epithelial and sarcomatous elements, and it is classified to be within homologous or heterologous types according to the origin of its mesenchymal tissue. Homologous MMMTcontains malignant stromal elements native to the ovary, whereas heterologous MMMT contains sarcomatous tissue not normally found in the ovary, such as bone or cartilage [4]. In one report, there was no statistical difference in survival between homologous and heterologous MMMT [5].

Research in malignant mixed mesodermal tumours ofthe uterus have suggested that the sarcomatous and carcinomatous components both arise from a single malignant epithelial precursor which has undergone metaplastic change to a sarcomatous form in some areas of the malignant tissue which contributed to the presence of both histological types [6]. Primary ovarian carcinosarcomas are very aggressive and are usually diagnosed at an advanced age and an advanced stage of disease [7]. The adverse prognostic factors as enumerated by various studies include advanced age, advanced stage, suboptimal cytoreduction, stromal predominant tumours and tumours with serous epithelial component [8]. The survival for both early and late stage carcinosarcomais inferior to serous tumours of the ovary [9]. The average survival for a woman diagnosed with carcinosarcoma of the ovary is <2 years [10].

Conclusions

To summarize, malignant, mixed mullerian tumors ofthe ovary are very aggressive tumors that were usually diagnosed at an older age compared to women with epithelial ovarian cancer. Similar to this one, MMMT are usuallyat an advanced stage at the time of diagnosis, and survival after diagnosis varies by stage of disease and histological type. Despite aggressive treatment that includes surgery and chemotherapy, women with these tumors have a significant increased risk of death compared to women with epithelial ovarian cancer and very poor prognosis [11]. The poor prognosis associated with this rare disease emphasizes the need for collaborative prospective studies targeted to better understand the molecular changes of MMMT and the need to design new therapeutic regimens to improve patients' survival.

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