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Case Report

A RARE CASE OF MALIGNANT MIXED MULLERIAN TUMOUR OF UTERINE CORPUS

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Abstract :
Malignant mixed mullerian tumors of the uterus are very rare neoplasms associated with an aggressive clinical course and overall poor prognosis. This tumour is having biphasic pattern consisting of both eithelial and mesenchymal components .We report a case of carcinosarcoma of the uterus successfully treated with surgery followed by chemotherapy.
Keywords: Carcinosarcoma,Chemotherapy, Malignant mixed mullerian tumor

INTRODUCTION

Uterine carcinosarcomas, also called malignant mixed mullerian tumors (MMMT), is a rare variant of cancer of reproductive tract, accounting for overall incidence of 1-4% [1].
These tumours are most commonly seen in postmenopausal women.MMMT is an aggressive tumour of uterine corpus.Poor prognosis is noted in advanced stage and with myometrial invasion .The Histologically, carcinosarcoma tumors are composed of both carcinomatous and mesenchymal components, which may be either homologous (composed of tissues normally found in the uterus) or heterologous (containing tissues which are not normally found in the uterus). They are aggressive and have a poor overall survival rate [2,3]. We report a case of carcinosarcoma of the uterus successfully treated with surgery(TAH +BSO+Pelvic lymphadenectomy) followed by chemotherapy with paclitaxel and cisplatin.

CASE REPORT

A 71-year-old woman who had history of weight loss of 10 kg in 1 year, 4 months of progressive diffuse dull aching abdominal pain with vaginal discharge and abdominal distension presented at our department of OBGY in july 2013.
Menstrual history revealed she was menopausal since 20 years.
Obstetric history suggested she was P7L7 ; All 7 full term vaginal deliveries at home.
Physical examination showed a pelvic mass. Pelvic ultrasound revealed large intramural anterior wall fibroid with small complex ovarian cyst on left side.The CA 125 levels were increased.The patient underwent suboptimal debulking surgery including total hysterectomy, bilateral salpingo-oophorectomy, omentectomy and biopsy of peritoneal node dissection. Gross examination of uterus showed fleshy ,friable mass in fundal region of uterus with areas of necrosis and haemorrhage within it. Adjuvant therapy (cisplatinum and paclitaxel) was also included in treatment program following surgery.
The pathologic report was uterine MMMT . Histologically, the major part was sarcomatous poorly differentiated with the carcinomatous part. Disease stage was Ia uterine MMMT according to the criteria of the international Federation of Gynecology and Obstetrics.

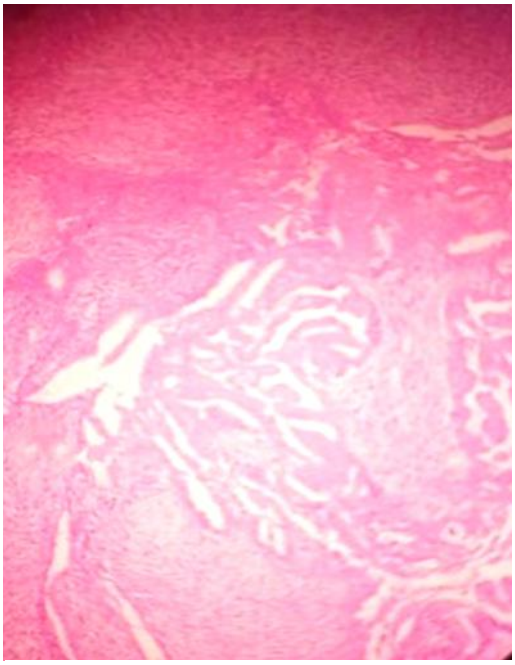

Fig 1 MMMT of uterus showing both carcinomatous and sarcomatous component.

DISCUSSION

Malignant mixed mullerian tumors(MMMT) of the uterus are extremely rare neoplasms that are most commonly seen in postmenopausal patients.Apart from uterine corpus MMMT are also found in cervix,ovaries,fallopian tubes,vagina,peritonium and also extragenital sites They mostly present with uterine bleeding,pelvic mass and pain abdomen. The usual location in uterus is the uterine body, particularly the posterior wall of the fundus but a few cases with MMMT of the uterine cervix have been reported as well[4]. The predisposing factors for MMMT include nulliparity,diabetes, obesity,chronic estrogen stimulation and history of pelvic radiation therapy in the past.Cases of tamoxifen associated uterine MMMT also have been reported.[5]

Grossly, these tumors present as large, soft, broad based and polypoid masses involving the endometrium and myometrium with fleshy surfaces. Necrosis and hemorrhage are commonly found as well. As the tumour is highly aggressive ,extensive myometrial invasion is usually seen.The characteristic microscopic features of MMMTs are the admixture of carcinomatous and sarcoma like elements. The carcinomatous component is usually a poorly differentiated adenocarcinoma. The most common sarcomatous components are homologous (endometrial stromal sarcoma, leiomyosarcoma) or heterologous(muscle,cartilage,osteoid,fat). Angiolymphatic invasion is very common.Rarely neuroectodermal differentiation is also seen.[6]

Due to aggressive nature and poor prognosis of MMMT,various therapeutic modalities have been employed.Surgical management includes total abdominal hysterectomy and bilteral salpingo-oophorectomy,infacolic omentectomy and bilteral pelvic and para-aortic lymphadenectomy.

There is no consensus on the optimal adjuvant chemotherapeutic regimen in uterine carcinosarcoma. Multiple chemotherapeutic regimens have been evaluated with modest response rates ranging from 12% to 100%. The trials are difficult to evaluate due to small numbers of patients, multiple treatment regimens, and occasional use of radiation therapy. The chemotherapeutic regimens most commonly utilized can be simplified into platinum

containing regimens compared to non-platinum regimens. A review of the literature does seem to support the use of platinum-based chemotherapy regimens with a 68% overall response rate in the platinum group compared with 23% response rate in the non-platinum containing regimens [7,8].

Based on uterine sarcoma data, many institutions combine ifosfamide with platinum, [9] while others follow more traditional epithelial ovarian regimens.

As this tumour is highly aggressive,prognosis is very bad.5 year disease free survival by stage is poor(stage I-56%,stage II-31%,stage III -13%, stage IV -0%) with most patients developing extrapelvic disease.[10].

To summarize, malignant, mixed mullerian tumors of the uterus are very aggressive tumors that were usually diagnosed at an older age. 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].

CONCLUSION:

Malignant, mixed mullerian tumors of the uterus are extremely rare and very aggressive tumors that were usually diagnosed at an older age. Clinician should have high index of suspicion and consider the possibility of MMMT of uterus. The outcome correlates with the stage of the disease and the depth of myometrial invasion .The poor prognosis largely depends on the stage at the time of diagnosis.With better understanding of the disease ,combination chemotherapy after radical surgery remains the mainstem of the treatment. However the optimal adjuvant treatment for this uncommon disease is yet to be established,indicating the need for larger multicentric studies on this tumour.

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