Malignant mixed Mullerian tumour (MMMT) of uterus: Rare and aggressive Tumor

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Introduction
A malignant mixed Mullerian tumour (MMMT), also known as uterine carcinosarcoma (UC). MMTT is an extremely rare and very aggressive tumour, comprising only 1–2% of uterine neoplasms1. In the United States, the annual incidence of uterine sarcomas is about 2 per 100,000 women; more than 50% of these are MMMTs. MMTT have a poorer prognosis than those with uterine carcinoma. Five-year survival rates of patients with MMTT that range between 33% and 39%.2 Most of women are post-menopause, especially between the 6th and the 7th decade.3 Risk factors of MMTT are nulliparity, obesity, advanced age, exposure to exogenous oestrogens, pelvic irradiation, and long-term use of tamoxifen.4,5,8 MMTT is responsible for 15% of deaths from uterine malignancy.7 More than 50% of MMTT patients present with advanced-stage disease.8 MMTT recently classified as high-grade endometrial carcinoma.9,10 MMTT are characterized by both a carcinomatous (eg, endometrioid, serous, and clear cell) and a sarcomatous component, and the sarcoma can contain either homologous (eg, fibrosarcoma, leiomyosarcoma) or heterologous, nonnative (eg, rhabdomyosarcoma and osteosarcoma) elements. Prognostic factors of MMTT are surgical stage, lymphovascular space invasion (LVSI), depth of myometrial invasion, tumor histology, and patient age.11 Since MMTTs are extremely rare and there is a paucity of data, we conducted the present study to analyse the outcome of this type of tumour. Here we report two cases of malignant mixed Mullerian tumor of the uterus with homologous elements.

Case Reports

1) A 70 years old female patient presented with chief complaint of Postmenopausal bleeding. CECT-Abdomen shows a large fluid filled collection with homogenous wall in pelvis. Total Abdominal Hystectomy (TAH) with Bilateral Salpingo Ophrectomy (BSO) was done. Post optrative Histopathological examination (HPE) shows malignant mixed Mullerian tumor (high grade (Image-1 & 2)) with Lympho Vascular Space Invasion (LVSI). Patient received adjuvant Radiotherapy 50Gy/25Fr/5 week to pelvis by Antero-Posterior(AP)/Postero-Anterior (PA) Field
f/b Supplementary Radiotherapy 16Gy/8Fr with reduced field size to pelvis. Instead of Brachytherapy by Cobalt 60 machine. Despite these efforts, patient developed Omental metastasis on follow up.

2) A 70 years old female patient presented with chief complaint of discharge P/V and postmenopausal bleeding. CECT-Abdomen shows enlarged uterus with endometrial thickening along with moderate ascites and minimal omental thickening. TAH with BSO was done. Post op HPE shows malignant mixed Mullerian tumor. One out of 20 pelvic LN show metastatic deposits. In view of poor general condition and advanced age she was not offered adjuvant therapy. Patient is kept on close follow up with symptomatic and supportive management.

Discussion of Treatment
The principal treatment for MMMT of the uterus is surgery, but the high rates of both local and distant relapse after surgery have demonstrated the need for effective adjuvant therapies. Surgery, including hysterectomy and bilateral salpingooopherectomy with surgical staging, is the mainstay of treatment of malignant mixed Mullerian tumour (MMMT). Indication of adjuvant radiation therapy for the treatment of carcinosarcoma are post-operative residual disease or surgical inaccessible sites as well as lymph node status. Postoperative External Beam Radiotherapy Therapy (EBRT) + Brachytherapy (BT) combination is associated with an overall survival advantage in malignant mixed Mullerian tumour (MMMT). External beam radiation therapy improves locoregional control in all stages. Radiation therapy to be given in doses of 50-60 Gy to the pelvis.

Conclusion
In conclusion, MMMTs are a rare and aggressive tumour of the uterus with poor prognosis so clinician should have a high index of suspicion of MMMT in postmenopausal patients and there is need to evaluate role of adjuvant radiation therapy for aggressive nature of tumor.

Reference: