

Case Report

Carcinosarcoma of the Uterus in a Patient with Previous History of Carcinoma of the Vulva

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ABSTRACT

Carcinosarcoma or Malignant Mixed Mullerian Tumour (MMMT) is a rare, but aggressive tumour of the post menopausal patients. Here we report one patient who

had undergone treatment for carcinoma of vulva and who later developed MMMT-heterologous type, for its unusual and rare occurrence.

KEYWORDS: carcinoma vulva, carcinosarcoma, malignant mixed mullerian tumour

INTRODUCTION

Carcinosarcoma popularly called as 'Malignant Mixed Mullerian Tumour' (MMMT) or 'Malignant Mixed Mesodermal Tumor', of the female genital tract is a rare, but aggressive neoplasm occurring more commonly in postmenopausal patients with low parity^[1]. The tumor arises in the tissues derived from the mullerian system and can occur in any of the pelvic structures covered by the celomic epithelium as well as from the genital tract derived from the mullerian duct. They contain both an epithelial and a mesenchymal component but the epithelial component seems to be the more predominant metastatic lesions^[1]. These tumors have a very poor prognosis irrespective of clinical stage, histological grade or the type of the components^[2]. There have been reports of other types of malignancy occurring along with MMMT but cases of MMMT following a previous malignancy elsewhere are extremely rare. We describe such an unusual and rare condition in a patient who had carcinoma of vulva followed by MMMT in the uterus 13 years later.

She was treated in this department and was given external radiation therapy by Cobalt 60 teletherapy to the vulva and bilateral inguino-femoral regions up to a tumor dose of 7000cGy/35 exposures in seven weeks. There was good regression of the lesion, with complete regression clinically.

Adjuvant chemotherapy was given for six cycles in the form of Cisplatin-30 mg, Etoposide 100 mg given from day 1-3, cyclophosphamide 500 mg on day 1, as a 3-weekly cycle. Then, after a period of six years of the treatment of the recurrence, 13 years after the carcinoma vulva, she reported to the department again with the aforesaid complaints. She was investigated and was found to have a

CASE REPORT

A 63-year-old, P₂ G₂, post-menopausal lady, attended the Department of Radiotherapy, Regional Institute of Medical Sciences, Imphal, with complaints of foul smelling vaginal discharge and occasional vaginal bleeding. Past history revealed that she had been operated (vulvectomy) 13 years ago for well differentiated squamous cell carcinoma of vulva. Seven years later, she developed a recurrence on the same site with ulceration and inguinal lymph node involvement.

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Fig. 1: Photomicrograph showing a section of uterus showing MMMT – both adenocarcinoma (arrowhead) and sarcoma (predominantly lipoblasts, thin arrow) components. (H & E stain, x 100)

uterus^[3]. The tumor can occur synchronously or metachronously with other ovarian, or pelvic tumours with similar or different histogenesis^[3]. The median age of such patients is about 65 years. There is little information in the literature on the metachronous association of the tumor with vulvar carcinoma, as was seen in our patient. Mayberg *et al* in 2000, reported a case of heterologous MMMT 12 years after the treatment of Hodgkin's disease by whole body irradiation^[4]. In the present case, MMMT developed in a shorter duration i.e., six years after the treatment with radiation and chemotherapy. Uterine MMMT usually presents earlier as compared to other extra-uterine MMMT; 15/25 patients presented in early stage^[1]. Histologically, the endometrioid carcinoma is seen predominantly as the carcinomatous component while the sarcoma component may be homologous or heterologous in cellular structure and usually graded according to their mitotic activity^[1,5]. Prognostically, the stage of the disease at presentation seems to be the only main prognostic factor with stage I patients having a five year survival of 62.3% while no patients beyond stage I survive up to five years^[1,6]. MMMT having endometrioid adenocarcinoma appears to have the most favorable prognostic factor besides the stage of the disease. Other prognostic factors appear to be the presence of heterologous component of the mesenchymal element, age of the patient and high grade tumor^[1,6].

One patient who had developed MMMT after undergoing pelvic radiation and surgery developed the tumor eight years after the treatment and died three months later^[1]. Our patient developed MMMT six years after having undergone radiation to vulvar and bilateral inguino-femoral regions followed by chemotherapy containing Cisplatin, Ifosfamide and Cyclophosphamide. The incidence of a MMMT after treatment of carcinoma vulva has not been described up to now in the literature. Whether radiation and/or chemotherapy is a factor in the development of MMMT is not very clear. The causal relationship between the two tumors, and radiation with chemotherapy for the previous tumor cannot be fully established. The origin of the tumor is still debatable but data or evidences available seem to suggest the concept of a metaplastic theory and a single stem cell origin^[1,3].

Though surgery is the main modality of treatment, adjuvant chemotherapy may have some role in the management of MMMT. Literature suggest that the chemotherapeutic agents Cisplatin, Ifosfamide, and Taxol have some effect on MMMT^[7]. The survival rate in such cases varies according to the mesenchymal component. For the homologous type the two and five year survival rates were 52% and 36% respectively, while 47% and 14% were seen respectively in the heterologous variety^[6].

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