

Malignant Mixed Mullerian Tumor of Uterus: Rare Case & Rarer Diagnosis

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Abstract

Carcinosarcoma or malignant mixed mullerian tumor (MMMT) of uterus is a rare tumor, representing only 1-2 % of all uterine neoplasm. The pre-operative diagnosis of MMMT can be a challenge as no preoperative diagnostic test including endometrial biopsy, CT or MRI has enough sensitivity or specificity. We are reporting one such case of MMMT where endometrial biopsy failed to make a pre-operative diagnosis to highlight the fact that a definitive pre-operative diagnosis can only be made in a small proportion of such cases thus precluding optimal surgery in such cases.

Keywords: Carcinosarcoma, Malignant mixed mullerian tumor, Endometrial biopsy.

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Introduction

Carcinosarcoma or malignant mixed mullerian tumor (MMMT) of uterus is a rare tumor, representing only 1-2 % of all uterine neoplasm^[1]. They are thus called because they contain a mixed composition of malignant epithelial and malignant mesenchymal components. Most commonly these tumors have a predilection for post-menopausal women but rarely are reported in younger women also. Clinically, patients with uterine MMMT present with abnormal vaginal bleeding.^[2,3] The pre-operative diagnosis of MMMT can be a challenge due to technical reasons leading to missed diagnosis and mismanagement. We are reporting one such case of MMMT where endometrial biopsy failed to make a pre-operative diagnosis to highlight the fact that a definitive pre-operative diagnosis can only be made in a small proportion of such cases thus precluding optimal surgery in such cases.

Case report

A 70yr old multiparous postmenopausal woman presented with postmenopausal bleeding for three weeks. There was no other noticeable history. Abdominal and speculum examination was unremarkable. Bimanual examination revealed an eight week sized soft uterus. Transvaginal ultrasound showed a large heterogenous echogenic lesion occupying entire uterine cavity with surrounding myometrium stretched and thinned out. CA-125 was 25 ng/ml. Endometrial

biopsy revealed necrotic tissue on histopathological examination. Total abdominal hysterectomy with bilateral salpingo oophorectomy was performed. On gross examination uterine cavity was distended by a well defined endometrial growth. Histopathology report revealed malignant mixed mullerian tumor stage 1A. Patient was advised adjuvant chemotherapy, however patient was lost to follow up. Patient was contacted telephonically at the time of writing this manuscript and was alive and healthy.

Discussion

Malignant mixed mullerian tumor represents the most common type of uterine sarcoma^[4]. There is a very little data about the clinical profile and outcome in literature. Carcinosarcomas comprise less than 5% of all uterine tumors however, it causes 16.4% of all deaths caused by a uterine malignancy.^[1]

Malignant Mixed Mullerian tumor (MMMT) is composed of a malignant epithelial component and a malignant stromal component. The epithelial component is typically a high-grade carcinoma of endometrioid or non-endometrioid type. There are divergent views about which epithelial component is more common, but it appears that the majority are endometrioid types^[5]. Similarly the stromal component may comprise homologous or heterologous tissue. When the stromal component produces tissue not normally found in the uterus, such as bone, cartilage, or skeletal muscle (osteosarcoma, chondrosarcoma, rhabdomyosarcoma), the tumors are designated as heterologous. A number of theories explaining the origin of MMT have been described in literature^[6] such as; that the carcinoma and sarcoma are two independent neoplasms (collision theory), that both components and derived from a single stem cell that undergoes divergent differentiation early in the evolution of the tumor (combination theory) and that, the sarcomatous elements derive from the carcinoma during the

evolution of the tumor (conversion theory). We could not make a diagnosis on uterine curettage and this is in keeping with the literature which suggest that a preoperative tissue diagnosis may only be possible in 20% to 50% cases.^[7,8] This is explained by a small amount of tissue obtained on endometrial biopsy. This small tissue may not be sufficient enough to identify both components and the epithelial component may be the only one recognized pre operatively. The diagnosis is most often made post hysterectomy on histopathological examination and immunohistochemistry(IHC). These tumors occur predominantly in postmenopausal women and generally are associated with a poor prognosis. The 5-year survival rates for FIGO stage 1 to 4 tumors are 45.8%, 30%, 10% and 0% respectively.^[9] Aggressive nature and poor prognosis of MMMTs necessitates aggressive management with various therapeutic modalities. At present recommended surgical treatment is total abdominal hysterectomy with bilateral salpingoophorectomy, infracolicomentectomy, pelvic lymphadenectomy and para-aortic lymph node sampling with peritoneal washings.^[10] However decision to do such aggressive surgery can only be made where a definitive pre-operative diagnosis is available, which is not the case in more than half of the cases. Adjuvant chemo radiation is recommended in most of the cases however there is no evidence that it offers any survival advantage.

MMMT are rare tumors and their accurate pre-operative diagnosis is even rarer. We believe that more efforts should be made to achieve an accurate pre-operative diagnosis in more number of cases, so that an optimal surgery may be performed. This might possibly lead to better survival.

Details of ethical approval: Not Required

Conflict of Interest: None

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