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Case Report

Mixed mullerian tumour presenting as giant uterine mass: a case report and review of literature

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ABSTRACT

Malignant mixed Müllerian tumors (MMMT) of the uterine corpus, also referred to as uterine carcinosarcomas, are rare and aggressive neoplasms, accounting for only 1–2% of uterine cancers. These tumors exhibit poor prognosis, with a 5-year survival rate of approximately 30% and only 50% being diagnosed at an early stage. Early detection is crucial, and imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) play a critical role in assessing tumor size, metastasis, and infiltration. The standard treatment involves surgical resection combined with platinum-based chemotherapy. Here, we present a rare case of a giant uterine mass in a 67-year-old female, confirmed to be MMMT. The patient underwent surgical debulking followed by chemotherapy, achieving significant resolution as evidenced by imaging. This case highlights the importance of early diagnosis and multimodal treatment in improving outcomes in MMMT.

Keywords: Malignant mixed Müllerian tumour, Giant uterine mass, Massive abdominal distension, Carcinosarcoma, Chemotherapy

INTRODUCTION

Malignant mixed Müllerian tumors (MMMT), or uterine carcinosarcomas, represent a rare subset of uterine malignancies, constituting only 1–2% of all uterine cancers. These biphasic tumors are composed of epithelial (carcinomatous) and mesenchymal (sarcomatous) elements, and they exhibit highly aggressive behavior with a propensity for early metastasis. The prognosis for MMMT remains poor, with advanced stages at diagnosis contributing to limited survival outcomes.[1] The clinical presentation of MMMT is variable but commonly includes postmenopausal vaginal bleeding, abdominal distension, or pelvic pain. Imaging techniques such as CT and MRI are pivotal in assessing the extent of disease and guiding management strategies. Surgical resection, typically involving hysterectomy with salpingo-oophorectomy, forms the cornerstone of treatment, with adjuvant chemotherapy, often using carboplatin and paclitaxel, improving survival rates.[2] Here, we report a unique case of MMMT presenting as a giant uterine mass in a 67-year-old female. This case emphasizes the challenges in diagnosis and the importance of a multimodal approach to management.

CASE REPORT

A 67-year-old post-menopausal female (Para 2+2) presented to emergency room with complaints of intermittent breathlessness since 3 months, increased over last 2 weeks associated with abdominal distension, bilateral pedal edema since 15 days. There was a history of weight gain since last 6 months. Patient attained

menopause at the age of 48 years. There was no history of post-menopausal vaginal bleeding. There was no family history of cancers. At admission, her SP02 was 80 % pulse was 112/min, BP was normal. Bilateral pedal edema and abdominal distension was present. CT abdomen-pelvis was suggestive of malignant complex cystic ovarian mass with metastatic disease, moderate ascites, paraumbilical hernia, tumor encasing the entire genital tract, bilateral pleural effusion. Hemoglobin was 7.8 gm/dl before surgery. Patient was optimized preop and underwent debulking surgery that included excision of large adnexal mass measuring sized 25×30 cm, 8 kg weight and adhesiolysis, hysterectomy, omentectomy was also done. Large size of tumor was compressing inferior vena cava hence decreased venous return along with pedal edema was present. Despite large size of tumor CA-125 was only 491.6 U/ml indicated possibility of non-epithelial origin before histopathology report.

Histopathology report revealed large tumour mass which was high grade malignant mixed mullerian tumor (carcinosarcoma)-primary from the outer uterine myometrium. Uterus showed cystic atrophy of endometrium. Both ovaries and fallopian tubes were free of tumor. Omentum and umbilical metastasis were also noted. Ascitic fluid was negative for malignant cells. Disease localized to abdomen with ascites and metastasis to omentum and peritoneum indicated clinical Stage III while pathological stage of tumor was pT3. Post operatively patient improved, compression symptoms resolved and was started on chemotherapy for malignancy. Patient underwent post-operative chemotherapy 1 month after operation, total 6 cycles of chemotherapy with Inj. Paclitaxel 300 mg and Inj. Carboplatin 600 mg along with supportive medications. Repeat 18 FDG-PET scan of whole body showed complete resolution with no residual masses after 6 cycles.

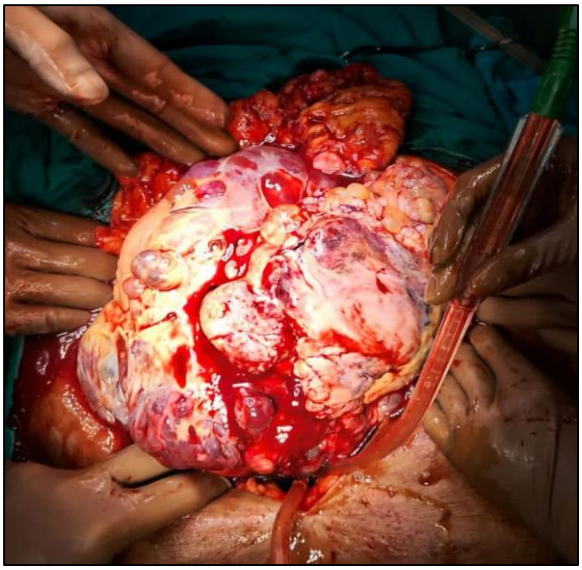

Figure 1: Intraoperative image of large adnexal mass of 8 kg weight and size 25×30 cm.

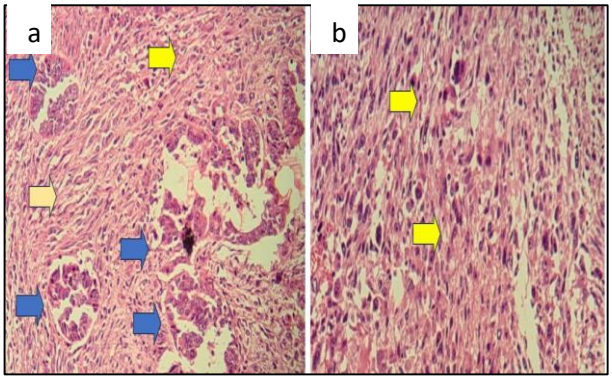

Figure 2: Histopathology-microphotography of carcinosarcoma showing (a) carcinoma component (Blue arrows) and (b) sarcomatous component (Yellow arrows).

DISCUSSION

Carcinosarcoma predominantly affects the uterus and is also referred to as malignant mixed müllerian tumor. However, it can also arise in the cervix, ovaries, fallopian tubes, vagina, peritoneum and extra-genital.[2] The majority of affected individuals present with postmenopausal vaginal bleeding, occasionally accompanied by a protuberant fleshy mass from the cervix. The clinical manifestation of uterine MMMT is contingent upon its anatomical origin. Most patients present with irregular vaginal bleeding which may range from mild to profuse. In this particular case the absence of vaginal bleeding was attributed to the confinement of the tumor to the outer uterine layer. Additional symptoms may include progressive fatigue, abdominal distension, pain, or a palpable abdominal mass, with some patients reporting increased abdominal girth. In advanced stages, involvement of the gastrointestinal or urinary tract may lead to related symptomatology.[3]

Carcinosarcoma typically presents in the fifth decade of life but is rarely observed in younger individuals. Studies suggest that postmenopausal women with low parity are at increased risk. The mean age of onset is approximately 65.5 years (range 55–77 years), and most cases exhibit disseminated disease at the time of diagnosis.[4] The prognosis is poor, particularly in individuals with extrauterine dissemination at the time of diagnosis.[5] The classical triad of symptoms indicative of MMMT includes pain, profuse vaginal bleeding, and the passage of necrotic tissue per vaginum.[6] The five-year survival rate is approximately 30% for stage III disease, whereas early-stage cases (stage I) have a survival rate of around 50%.[7]

Hypotheses with respect to the histogenesis of MMMT have been proposed, including collision, combination and conversion speculations. The collision hypothesis suggests that carcinoma and sarcoma are two autonomous neoplasms, whereas the combination hypothesis postulates that both originate from a common progenitor

cell undergoing divergent differentiation. The conversion hypothesis posits that the sarcomatous component evolves from the carcinomatous element during tumor progression. McCluggage proposed that the spindle cell component represents a pseudo sarcomatous stromal reaction to carcinoma with divergent differentiation. Immunohistochemical and molecular studies indicate that MMMT may originate as an adenocarcinoma, subsequently acquiring sarcomatoid differentiation over time, supporting the clonal evolution of these neoplasms. Both high-grade endometrial carcinoma and MMMT exhibit similar molecular profiles, with TP53 mutations being the most prevalent genetic alteration. However, immunohistochemistry is not essential for diagnosis.[8,9]

Therapeutic strategies for MMMT remain challenging due to its aggressive nature and poor prognosis. Although evidence is limited, expert recommendations support the use of platinum-based combination chemotherapy. Carboplatin and paclitaxel are the preferred regimen for patients with newly diagnosed uterine carcinosarcoma following surgical resection.[7] Surgical excision remains the cornerstone of management, with adjuvant chemotherapy and radiation therapy improving overall survival. Prognostic factors include patient age, tumor size at presentation, depth of myometrial invasion, and the presence of malignant cells in peritoneal cytology. Given that MMMT predominantly affects postmenopausal individuals with multiple comorbidities, treatment planning should be individualized to optimize outcomes.[10]

CONCLUSION

In conclusion, we have described uterine MMMT which is rare and demonstrate variable CT and MRI morphological appearances. Due to the heterogeneous nature and very high morbidity of MMMT, combination of careful analysis of imaging findings and clinical features might be useful for a more accurate diagnosis of MMMT.

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