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A Study of Two Uncommon Gynecological Malignancies

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ABSTRACT

In busy gynecological practice, we come across many rare and uncommon malignancies. Here we present two uncommon varieties, one, synchronous tumours of endometrium and ovary, and the other, Malignant Mixed Mullerian Tumours. Simultaneously occurring tumours in the endometrium and the ovary can be either primaries in both the organs or metastasis from one another. It is important to differentiate the two as the prognosis and management varies depending on whether it is primary or metastatic. Malignant Mixed Mullerian Tumours (MMMT), also called as uterine carcinosarcoma, is a rare and rapidly growing tumour of the female genital tract.

Introduction

In busy gynecological practice, we come across many rare and uncommon malignancies. Here we present two uncommon varieties, one, synchronous tumours of endometrium and ovary, and the other, Malignant Mixed Mullerian Tumours. Malignant Mixed Mullerian Tumours (MMMT), also called as uterine carcinosarcoma, is a rare and rapidly growing tumour of the female genital tract.[1] The occurrence of simultaneous primary cancers, that is, synchronous cancers in the endometrium and ovary is uncommon.[2] Most of the reported cases have tumours of similar histology. Here we present a case of synchronous endometrial and ovarian cancer with dissimilar histology.

Synchronous Carcinoma Ovary with Carcinoma Endometrium

Introduction

Simultaneously occurring tumours in the endometrium and the ovary can be either primaries in both the organs or metastasis from one another. It is important to differentiate the two as the prognosis and management varies depending on whether it is primary or metastatic. These tumours occur synchronously as the surface epithelium of the ovary and the mullerian system has the same embryonic development.[3]

Case Report

A 55 year old, postmenopausal nulliparous patient was admitted in our gynecology ward with history of gradual distension of abdomen followed by pain in abdomen over a period of 3 months. There was a history of loss of weight and loss of appetite. USG and MRI confirmed the presence of a large mass 15.6x9.2x13 cm with numerous peritoneal deposits largest 3.1 cm in size. There was also evidence of polypoidal masses in the endometrial cavity. She was assigned clinically as Stage IIIC. CA-125 levels done at that time were 807.3 U/ml. In view of the advanced stage of the ovarian malignancy decision was taken to give Neo-adjuvant chemotherapy followed by surgery. FNAC of the mass was done which confirmed it as a case of papillary serous cyst adeno carcinoma. Patient was given two cycles of Neo-adjuvant Chemotherapy with Carboplatin and Paclitaxel following which she underwent total abdominal hysterectomy with bilateral salpingo-ovariotomy with omentectomy. Histo-pathological examination confirmed the presence of papillary serous cyst adeno carcinoma of the ovary with synchronous endometriod carcinoma of the endometrium involving the inner half of the myometrium. She was given 4 more cycles of chemotherapy but subsequently was lost to follow up.

Discussion

Around 10% of all women with ovarian cancer have a coexistent endometrial cancer and 5% of all women with endometrial cancer have a coexisting ovarian cancer. i These cancers may be mistaken as FIGO Stage III of endometrial cancer or FIGO Stage II of ovarian cancer. [4] Women with synchronous tumours present by the fifth decade and have an improved survival. i Pearl et al[5] however reported 63% incidence in the post menopausal age group like our patient. The presenting complaints are abnormal uterine bleeding (AUB), abdominal mass, abdominal pain and abdominal fullness. i Our patient presented with abdominal distension with abdominal pain. Elevated CA-125 levels i are sometimes present as in our patient. Yamanoi et al [6] reported that 46% of the patients were nullipara as in our case. Ulbright and Roth [7] proposed pathologic criteria for distinguishing between synchronous and metastatic tumours in 1985. These were further refined by Young and Scully.[8]Immuno-histochemistry has been showed to be useful in identifying primary from metastasis.[9] Ayhan et al [10] reported that prognosis depends on stage of the ovarian tumour and grade of the endometrial tumour. Zaino et al [iii] and Chiang et al [11] reported that stage of the tumour is more important in survival. Surgery is the mainstay of treatment in these tumours. x Appropriate post operative chemotherapy and or radiotherapy should be given as per the stage at which the diagnosis is made. These patients may not require further radiotherapy or chemotherapy as they usually present at an early stage. [viii] Pearl et al, [iv] reported a survival of 100% in these women. Castro et al [12] reported that though these patients present at a late age, they have a good survival as they have an early stage disease. Atallah et al [13] reported a 25 year old nulliparous patient with a synchronous tumour who had a full term spontaneous pregnancy after a conservative surgery and adjuvant chemotherapy with hormonal treatment.

Conclusion:

Women with synchronous dual primary ovarian and endometrial carcinomas have a better outcome than the women with a single cancer in either of the organs. Proper guidelines are needed for staging and treatment of these tumours.

Malignant Mixed Mullerian Tumours

Introduction

MMMT may be seen in vulva, vagina, cervix, endometrium (most common), ovary or fallopian tube. [14] The theories of origin of these tumours are: 1.Biclonal theory- which says that both the tumours arise independently 2. Monoclonal theory- which says that they arise from a single neoplasm and then undergo multidirectional differentiation. [xv] A study of 43 MMMT cases favoured monoclonal theory.[xiv] We present a case of MMMT coming to our institution.

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Case report

A 60 year old, Para 4, postmenopausal patient, came with history of postmenopausal bleeding for 2 and half months. She was a known case of diabetes mellitus and hypertension on regular treatment for 4 years. On examination, vitals were stable. On per abdomen examination, there was a mass arising from pelvis, hard in consistency, corresponding to 16 to 18 weeks pregnant uterus. On per speculum examination, there was a necrotic mass coming out of the cervical os. On per vaginal examination, same findings were confirmed.

Investigations

Hb=11.9gm%; TC=7500/cmm; DC=55/36/5/3/1; FB/PP=148/180mg%; RFT/LFT=WNL; CA 125=42.5U/ml

USG and MRI pelvis confirmed the findings of enlarged uterus with a large multicentric mass in the endometrial cavity. Both ovaries were normal. With a preoperative diagnosis of uterine malignancy, patient was taken up for surgery. Total Abdominal Extrafascial Hysterectomy with Bilateral Salphingo Oophorectomy was performed. Histopathological examination showed Malignant Mixed Mullerian Tumour infiltrating into 2/3rd of the myometrium. Patient was sent for post operative adjuvant radiotherapy.

Discussion

Malignant Mixed Mullerian Tumours (MMMT), also called as uterine carcinosarcoma, is a rare and rapidly growing tumour of the female genital tract. [xiii] They contain carcinomatous and sarcomatous elements. The sarcomatous element can be homologous or heterologous. Homologous type can be a spindle cell sarcoma, endometrial stromal sarcoma, leiomyosarcoma, malignant fibrous histiocytoma, undifferentiated sarcoma, or mixed. Heterologous type can be rhabdomyosarcoma, chondrosarcoma, osteosarcoma and liposarcoma. [i] These tumours are seen mainly in the postmenopausal age group as in our patient.[15] The symptom triad indicative of MMMT includes pain, severe vaginal bleeding and passage of necrotic tissue per vaginum.i Endometrial biopsy is not a good diagnostic modality for these tumours as shown by Rajshekar et al.[xvi] The most important prognostic features are the stage, the size of the tumor, and the depth of myometrial invasion.i Total abdominal hysterectomy with bilateral salphingo-oophorectomy, infracolic omentectomy, and bilateral pelvic and para-aortic lymphadenectomy is the primary mode of treatment.[xvi] Effective adjuvant therapy is still not outlined.[16] Paclitaxel and carboplatin is preferred as adjuvant chemotherapy.[17]

Conclusion

MMMT are rare and rapidly growing uterine cancers. As these are rare, definitive therapeutic guidelines have not been well outlined.

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