

CASE REPORT

MALIGNANT MIXED MULLERIAN TUMOUR, HETEROLOGOUS IN A 66-YEAR-OLD MALAY LADY - A CASE REPORT.

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A 66-year-old Malay woman, known hypertensive, presented with post menopausal bleeding associated with clot for three months. She was postmenopausal for last ten years. She also complaints of developing a mass in the abdomen which was growing in size also for last three months. Abdominal examination revealed a twenty week size mass, movable from side to side but unable to get below the mass. Vaginal examination revealed a fleshy fungating mass arising from the uterus coming out through the vagina. Cervix could not be visualized properly. Subsequent histopathology of the removed mass was reported as a Malignant Mixed Mullerian Tumour – Heterologous.

Key words : Malignant Mixed Mullerian Tumour, Heterologous.

Submitted-30.7.2002, Revised-11.6.2003, Accepted-15.6.2003

Introduction

Malignant Mixed Mullerian Tumour is a rare uterine neoplasm that is seen always in postmenopausal woman although exceptions occur (1). They present with uterine bleeding and enlargement and the usual location is uterine body. Grossly they present as large, soft, polypoid growth involving the endomyometrium and some times protruding through the cervix. Here we present an interesting case of Malignant Mixed Mullerian Tumor in a 66 year old Malay Lady.

Case History

The patient was a 66 year old Malay Woman who was a known hypertensive and was on Nifedipine 15mg TDS. She was admitted in the USM hospital on 22nd June 2002 with the complaints of postmenopausal bleeding associated with clot for last 3 months. She also complained of an abdominal mass increasing in size for the same three months. She had foul smelling vaginal discharge and also had history of loss of appetite and loss of weight.

Menstrual history

She attained menarche at the age of 13 years and had regular cycle for 7 days flow with 30 days cycle. She never had Pap's smear test. She never used any contraceptive device or pill before. She is still sexually active.

Obstetric history

Para 8. All of the deliveries were full term spontaneous vaginal deliveries except the last childbirth, which was a Caesarian section with bilateral tubal ligation. Physical examination revealed she was moderate built, had mild pallor and not cachectic. Her blood pressure was 200/100 mm Hg., temperature 38° C, CVS and RS were normal. Per abdomen examination showed a mass 20 weeks in size, firm, mobile from side to side, unable to get below the mass. There was no ascitis. Per vaginal examination revealed atrophic vagina and no other gross abnormality was detected in the vulvovagina area. An irregular fleshy fungating mass, arising from the uterus was seen. There was contact bleeding and foul smelling discharge. The Cervix could not be visualized. The adnexae and

pouch of Douglas were normal. Trans abdominal scan showed uterus grossly enlarged with discrete mass and areas of calcification. The clinical impression was an infected degenerative fibroid and to rule out endometrial carcinoma. She was treated with antihypertensive, I.V antibiotics (Zinacef+Gentamicin+Flagyl). Biopsy was done and the report came as infarcted malignant tumour. Subsequently she was planned for total hysterectomy with bilateral salpingo- oophorectomy (TAHBSO) and the operation was done on 26/06/02. The specimen was sent for histopathological examination.

Pathological Examination:

Gross examination:

Specimen consists of a total abdominal hysterectomy with bilateral salpingo- oophorectomy specimen (TAHBSO) which was sent already cut open and completely distorted measuring 110X120X60mm. Right fallopian tube measured 50X10mm, Right ovary measured 25X15X12mm. Left fallopian tube measured 50X10mm and the left ovary measured 20X15X5mm. The whole specimen weighed 900gm. The endometrial cavity was filled with grey white necrotic friable tumour. Myometrium measured 15mm in thickness. The tumor appeared to be confined to the endometrial cavity grossly. The cervix looks normal grossly. Seven blocks one each from the anterior and posterior lip of cervix, right ovary and tube, left ovary and tube and three blocks from the tumour with myometrium, one from the tumour proper were embedded.

Microscopic examination:

Multiples sections show a tumour in the uterine cavity. The tumour is an admixture of both carcinomatous and sarcomatous elements.(Fig-1). The carcinomatous element is composed of moderate to poorly differentiated endometrial glands of endometroid type. In few foci squamous metaplasia are also seen. The sarcomatous component is made up of anaplastic spindle shaped cells and large plump cells having large hyperchromatic nuclei with prominent nucleoli and moderate amount of pink cytoplasm. Some large myoblasts exhibiting cross striations were seen. In few areas the stroma is myxoid. Mitotic figures are

more than 15/ HPF in some foci.(Fig2)The tumour has involved 3/4th of the myometrium wall. Extensive areas of necrosis are seen. Sections from the cervix show chronic cervicitis. Sections from both ovaries and tubes are within normal limit. Special stains (IHC) for Cytokeratin, Vimentin, Desmin and Myoglobin (Fig-3) were positive. Dx: Malignant Mixed Mullerian Tumour, heterologous.

Discussion

Our patient presented with postmenopausal bleeding with passage of clot for last three months and also gradual abdominal swelling for the same periods. She also complained of foul smelling discharge from vagina. These all coincides with the usual presentation of Malignant Mixed Mullerian Tumour. Sarcomas collectively made up about 5% or less of uterine tumours, the commonest variant being mixed mesodermal tumors, leiomyosarcomas, endometrial stromal sarcomas (2). Malignant mixed Mullerian tumor is called carcinosarcomas because they consists of both glandular and stromal elements. The present case also showed carcinosarcoma in histopathology. The stromal components may differentiate into a variety of malignant mesodermal components like muscle, cartilage and bone ; malignant mixed Mullerian tumors are highly aggressive. The case we report here also showed rhabdomyoblasts in light microscopy where cross striation was easily identifiable and Immunohistochemistry(IHC) showed positivity for myoglobin and also for Desmin and Vimentin. In recent years convincing evidence suggested that most not all are monoclonal in origin rather than true collision tumors (3). Various data confirms that the sarcomatous component is derived from the carcinoma or from a stem cell that undergoes divergent differentiation. Thus uterine carcinosarcomas are best regarded as metaplastic carcinomas. Although Malignant Mixed Mullerian tumours are rare neoplasm's occurring mostly in uterus, rare cases of malignant mixed mullerian tumours have also been noted in the cervix (4,5,6), and the omentum (7). The lesion in the cervix was a poorly differentiated lesion composed of poorly differentiated epithelial component (cytokeratin positive), and a spindle cell component (vimentin positive) with heterologous (myeoblastic) differentiation. Also there was a report of primary peritoneal Mullerian adenosarcoma with sarcomatous overgrowth associated with endometriosis in a 50-year-old female (8).

Histologically the tumor was composed of benign mullerian glands and a sarcomatous stroma. Multiple foci of endometriosis were associated with pelvic mass. Both estrogen and progesterone receptors differ in function and expression with different neoplastic states. Recent studies has shown that Malignant Mixed Mullerian Tumours and Endometrial adenocarcinomas exhibit decreased ER alpha expression and significant loss of PR protein (9). We also did the ER & PR staining on the histological slides and these were negative. A recent study which studied clinical and pathological analysis of 106 cases (10) of uterine sarcoma showed that the malignant mixed mullerian tumour presented in 15.1% of the total cases, while the predominant sarcoma was leiomyosarcoma (63.2%), malignant endometrial interstitial sarcomas (21.7%). They noticed that the patients with leiomyosarcoma and malignant endometrial stromal sarcoma were younger and less than 50 years of age (70.1% and 60.9% respectively), while most of the malignant mixed mullerian tumour patient were above 50 years of age. The presenting complaints of these patients were abnormal vaginal bleeding (67.0%), palpable lower abdominal mass (32.1%), vaginal discharge (27.4%), and discomfort feeling (28.3%). They concluded that the clinical symptoms of uterine sarcoma are non specific (mostly abnormal vaginal bleeding) and the prognosis was poor. The prognosis of uterine sarcoma is related to histopathological sub types, clinical stages and age of the patient. The mainstay of treatment is surgery with adjuvant chemotherapy and radiotherapy.

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