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Case Report

A RARE CASE REPORT: MALIGNANT MIXED MULLERIAN TUMOR

Margaret TheresaJ[1*], Gerard Rakesh J[2], Balaji S[3], Fathima Jackia Banu I[3]

[1]Assistant Professor, Department of Pathology, Sri Venkateshwaraa Medical College Hospital and Research Centre, Pondicherry, India.
[2]Associate Professor, Department of Microbiology, Sri Venkateshwaraa Medical College Hospital and Research Centre, Pondicherry, India.
[3]Post graduate students, Department of Pathology, Sri Venkateshwaraa Medical College Hospital and Research Centre, Pondicherry, India

ABSTRACT

Malignant mixed mullerian tumor (MMMT) of uterus accounts <5% of tumor that arise from the uterine corpus. Highly aggressive and biphasic neoplasm composed of both epithelial and mesenchymal component with poor prognosis. We reported a case of 65 years old female presented with complaints of abdominal pain with vaginal bleeding. Routine investigations were within normal range. Radiological examination showed a large polypoidal mass arising in the endometrial cavity and extending upto the vagina. Total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH-BSO) was done, along with pelvic nodes were resected and the tumor was diagnosed as Malignant mixed mullerian tumor (carcinosarcoma)-Heterologous type of the uterus.

Key words: Malignant mixed mullerian tumor, Biphasic neoplasm, Carcinosarcoma.

Corresponding Author: **Margaret Theresa. J**
Email:margitheresa@gmail.com

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INTRODUCTION

Malignant mixed mullerian tumor(MMMT) of uterus accounts <5% of tumor that arise from the uterine corpus[1]. Synonyms are carcinosarcoma and metaplastic carcinoma. MMMT common among postmenopausal women, which is a highly aggressive neoplasm with poor prognosis. MMMT is a biphasic tumor which exhibit both epithelial and mesenchymal component and is divided into homologous type and heterologous type[2].

CASE REPORT

A 65 year old postmenopausal women came with the complaints of irregular and heavy bleeding per vaginum for past 2-4 months and history of abdominal pain and tenderness since 8 months. On general examination vitals were stable. Per speculum and per vaginal examination revealed a polypoidal mass protruding out of the cervix with mild tenderness. Bilateral fornices were free. Radiological examination showed a large polypoidal mass arising in the endometrial cavity, protruding out of the cervix and extending upto the vagina. Also seen are multiple fibroid in the myometrium. Total abdominal hysterectomy and bilateral salpingo-oophorectomy was done along with resection of pelvic nodes. Received TAH-BSO specimen with pelvic nodes. Uterus with cervix measuring 9x8x4.5cm. Cervix was hypertrophied. Cut surface of uterus shows a large polypoidal growth in the endometrial cavity, attached to fundus measuring 5.5x4x1.5cm, which is grey white to grey brown in color and firm in consistency. [Figure 1&2]. There is no obvious myometrial invasion. Cut surface of myometrium shows multiple fibroids. Largest measuring 2x2x1cm and smallest fibroid measuring 1x0.5x0.5cm.Cut surface is grey white, firm in consistency. Right ovary measuring 3x1.8x1cm, right fallopian tube measuring 3cm in length which are unremarkable. Left ovary measuring 3.5x1.8x1cm and left fallopian tube measuring 3cm in length are unremarkable. Also received right pelvic lymph nodes which showed grey yellow, fibrous fatty tissue fragments measuring 4x2x1cm .Cut surface shows five lymph nodes. Received left pelvic lymph nodes showed

single grey yellow, soft tissue fragment measuring 4x3x2cm.Cut surface shows three lymph nodes.

On Microscopic examination section studied shows a malignant neoplasm composed of both epithelial and mesenchymal component. The tumor cells from the epithelial component are arranged in sheets. The individual tumor cells are pleomorphic which exhibits round to oval shaped nucleus with increased nuclear cytoplasmic ratio with moderate amount of eosinophilic cytoplasm [Figure3]. Tumor from the mesenchymal component exhibit malignant chondrocytes and chondroblasts [Figure 4].Also seen are atypical mitotic figures with extensive areas of necrosis. No myometrial invasion identified.

Based on these findings, the histopathological diagnosis given was Malignant Mixed Mullerian tumor (carcinosarcoma)-Heterologous Type. Tumor-node-metastasis (TNM) staging was done T1aN0Mx Stage IA and the International Federation of Gynecology Obstetrics (FIGO) Stage IA. (T1a: Tumor limited to endometrium or invading less than half of myometrium. N0: N0 Regional lymph node Metastasis. Mx: Distant metastasis cannot be assessed.

Figure 1: Gross Finding Shows A Large Polypoidal Growth In The Endometrial Cavity

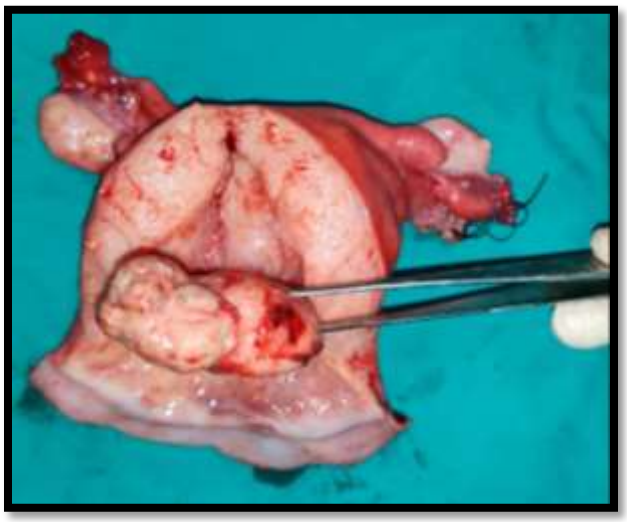


Figure 2: Gross Finding Show A Large Polypoidal Growth Extending Out Of The Cervix

Figure 3: Microscopy showing the epithelial component of tumor

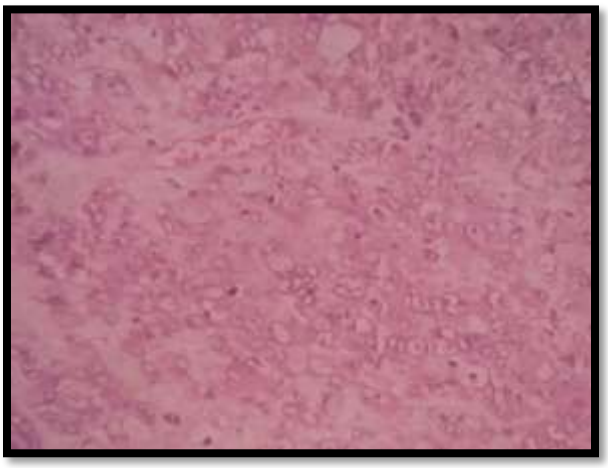
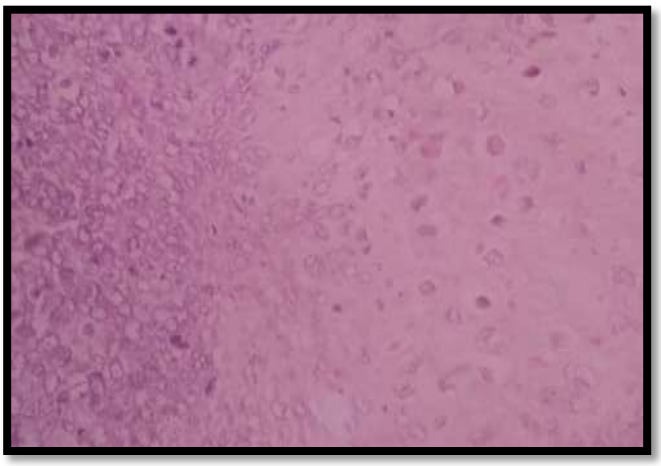


Figure 4: Microscopy showing the tumor composed of malignant chondrocytes and chondroblasts



DISCUSSION

Malignant mixed mullerian tumor is a rare, aggressive tumor of the female genital tract with an incidence of about < 2/ 10,000 women per year.[3] According to WHO this tumor falls under mixed epithelial and mesenchymal tumor type, hence it is also called as carcinosarcoma. The tumor commonly occur in the fifth decade of life. But at rare occasion it can occur in younger age as well, the lowest age of incidence was noted at 15-17years.[4] In the present study the age of presentation of this tumor is 65-year-old female.

Malignant mixed mullerian tumor are commonly found in uterine corpus, particularly in the posterior wall of the fundus but can be found in various other site like vagina, cervix, ovaries, fallopian tubes, peritoneum, and extragenital sites.[5] In our present study the tumor was found in the uterine corpus. Some of the risk factors that can trigger the development of Malignant mixed mullerian tumor are obesity, diabetes, prolonged estrogen stimulation, nulliparity and history of radiation in pelvic region. The common clinical presentations are lower abdominal pain, severe vaginal bleeding and abdominal distension.[6] In our case patient presented with complaints of irregular and heavy bleeding per vaginum, abdominal lower abdominal pain and tenderness.

On Gross examination, the tumors present as large, polypoidal mass with broad base. The tumor arising from endometrium with or without myometrial invasion. The tumor is soft in consistency and on cut surface tumor is fleshy with areas of hemorrhage and necrosis.[7] In the present study we observed a large polypoidal growth in the endometrial cavity attached to fundus, firm in consistency with focal areas of necrosis. On microscopy it is a biphasic neoplasm composed of both epithelial and mesenchymal components. Common epithelial component is adenocarcinoma, rarely mucinous, clear cell, serous and papillary serous components can also occur. Mesenchymal component is further classified into homologous and heterologous type according to the presence of sarcomatous component. In homologous type, the sarcomatous component is mainly composed of endometrial stroma and the smooth muscle or the fibrous tissue. In heterologous type is mainly composed of cartilage, bone and /or skeletal muscle.[8] In the present study we encountered a case of Malignant mixed mullerian tumor with heterologous type.

Malignant mixed mullerian tumor show evidence of metastasis through haematogenous route, lymphatic channels and direct extension into pelvic cavity. Recurrences are possible in half of the patients even after the surgical management and adjuvant therapy. The most common route is haematogenous. MMMT is managed surgically; Total abdominal hysterectomy-bilateral salpingo-oophorectomy with infra-colicomentectomy, bilateral pelvic and para-aortic lymphadenectomy is commonly indicated.[7] Multiple chemotherapeutic regimen also been suggested in various studies. In the present study there is no evidence of distance or lymph node metastasis and the surgical management done was Total abdominal hysterectomy with bilateral salpingo-oophorectomy along with pelvic lymph node resection.

Immunohistochemical marker for Malignant mixed mullerian tumor are epithelial membrane antigen and pancytokeratin which is the commonly used epithelial marker. Desmin and S-100 are the mesenchymal marker. However in the present study the microscopic features showed a clear evidence of Malignant mixed mullerian tumors with heterologous type.

CONCLUSION

Malignant mixed mullerianis tumor is a rare, aggressive tumor of the female genital tract which has a poor prognosis. Elderly female with complaints of irregular vaginal bleeding should be investigated in view of uterine malignancy and should also consider the possibility of Malignant mixed mullerian tumors. Surgery and chemotherapy is the mainstay of management. Prognosis of the disease is based on the stage and metastasis status of tumor.

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CONFLICT OF INTEREST

No conflicts of interest exist.

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