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CASE REPORT

A RARE CASE OF MALIGNANT MIXED MULLERIAN TUMOUR OF CERVIX – A CASE REPORT

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ABSTRACT

Background: Malignant mixed Mullerian tumors of the cervix (Carcinosarcomas or MMMTs) are rare and aggressive malignancies consisting of an epithelial (carcinoma) and a mesenchymal (sarcoma) tumor component.
Objective: To report a rare case of cervical malignant mixed Mulleriantumour.
Case: Here, we present a case of old postmenopausal woman with complaints of bleeding per vaginum, underwent endocervical curettage and endometrial biopsy suspicious of malignancy. So, radical hysterectomy with infracolic omentectomy and pelvic lymphadenectomy was performed and was later, diagnosed as malignant mixed Mulleriantumour arising from the cervix.
Conclusion: Because of the rarity of MMMT, there are no consensus guidelines regarding treatment. While hysterectomy with bilateral salpingo-oophorectomy remains the mainstay of treatment, high rates of recurrence, and metastasis suggests a need for lymphadenectomy and post operative adjuvant treatment

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INTRODUCTION

Malignant mixed Mulleriantumours (MMMT), first described by Ferrieraand colleagues in 1951are rare biphasic malignant tumour, affecting female genital tract (Soslow, 2009). MMMTs of cervix are extremely rare malignancies, constituting approximately 0.005% of all cervical malignancies. So far, only around 50 cases of cervical MMMTs have been reported in the literature (Maheshwari *et al.,* 2006; Kudela *et al.,* 2014). In the present report, we present a case of MMMT of cervix, in relation to clinic opathological profile.

CASE REPORT

A 70 year old, postmenopausal, obeselady presented with bleeding per vaginum for 5-6 months. She was a known diabetic, hypertensive with coronary artery disease& hypothyroid, controlled on treatment. Examination revealed enlarged uterus of 10 weeks size. Ultrasonography showed hyperechoic mass (9.6×2.4cm), filling the uterine cavity with increased vascularity of anterior myometrium. Endometrial biopsy and endocervical curettage was performed and showed poorly differentiated carcinoma. CT scan showed endometrial collection with differential enhancement of anterior and posterior lips of cervix, with no change in contour.

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Exploratory laparotomy was performed, which revealed enlarged uterus, calcified deposits in broad ligaments and infundibulopelvic ligaments and enlarged internal iliac and obturator lymph nodes. So, radical hysterectomy, with infracolicomentectomy and pelvic lymphadenectomy was done. Her postoperative recovery was uneventful. She was advised for radiotherapy and patient is under regular follow up for 4 months.

Pathological findings

Grossly, uterus with cervix, measured 10 x 6 x 8 cm. Cut surface showed dilated uterine cavity filled with friable polypoidal mass measuring, 8.5x5cm (Figure 1), arising from endocervix atsquamocolumnar junction. Endomyometrial thickness was 1cm. Light microscopy: Predominant papillaroidfrond like growth pattern was seen.Growth pattern was surrounded by undifferentiated anaplastic cells in abundance. Malignant cell types included clear cell, adenoid cystic, palisaded squamous, micropapillary and papillary pattern. Scanty scattered mesenchymal component composed of atypicalstrap cells and osteoid cells were identified, with large areas of coagulative necrosis. Endometrium and superficial 1/3 of myometrium was involved with involvement of bilateral obturator nodes and right common iliac nodes. The final diagnosis was malignant mixed Mullerian tumor of cervix stage III_B.

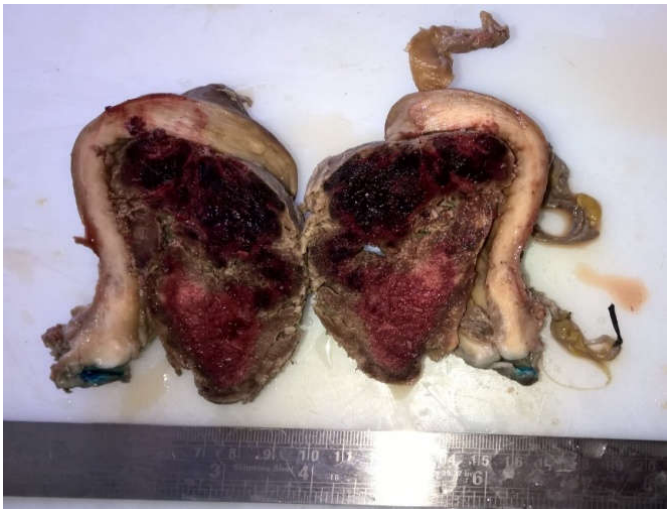
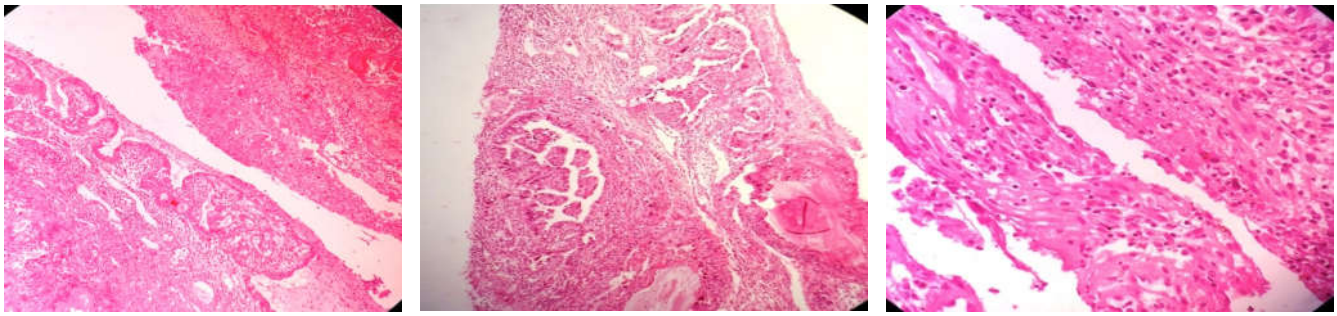


Fig. 1. Gross specimen



A.(H& E stain- 10X) **B. (H& E stain- 10X)** **C. (H& E stain- 40 X)**

Fig. 2(A-C). Histological sections show malignant epithelial component (adenocarcinoma) admixed with homologous stromal sarcomatous component and areas of hemorrhage

DISCUSSION

Cervical MMMTs are rare tumours, occurring usually in postmenopausal women, although age group varies from 12- 93 years. Most commonly patients present with abdominal or pelvic pain, vaginal bleeding, cervical growth or abnormal cytology (Maheshwari *et al.,* 2006). In our case, patient was 70 year old postmenopausal lady presenting with vaginal bleeding. Aetiological factors believed to be implicated in this cancer include exposure to irradiation, obesity, nulliparity, human papilloma virus or exogenous estrogen and long term use of tamoxifen (Kuyumcuoğlu *et al.,* 2009). In our case, patient was obese, with hypertension and diabetes. MMMT is a biphasic tumour arising in the female genital tract, composed of epithelial and mesenchymal tissues (Kuyumcuoğlu *et al.,* 2009). It is a rare malignant tumour, comprising 2-5% of all gynaecological malignancies. It mostly arises in uterus, less commonly in vagina, cervix, ovary, and rarely fallopian tubes. Based on sarcomatous component, two categories of carcinosarcoma of cervix have been identified: homologous andheterologous. The homologous type has a sarcomatouselements that are normal components of mullerian system like smooth muscle or fibroblasts –leiomyosarcoma, fibrosarcoma, while heterologous type contains cartilage, bone or skeletal muscles cells, foreign to affected site, like chondrosarcoma, osteosarcoma and rhabdomyosarcoma. In both cases, the carcinomatous component includes squamous cell carcinoma, basaloid squamous carcinoma, adenocarcinoma, adeno-squamous carcinoma, adenoid-basal carcinoma, adenoid-cystic carcinoma and undifferentiated carcinoma (Shim *et al.,* 2012). The pathological staging and histological features of the carcinomatous component of carcinosarcoma are responsible for the tumour's biological potential and aggressiveness. MMMTs express epithelial (EMA, Pancytokeratin) & stromal lineage marker in relation to their histological appearances such as vimentin, desmin in muscular differentiation or S100 in areas with chondroid or lipomatous differentiation (Shim *et al.,* 2012). However IHC studies are not mandatory for diagnosis of MMMTs.

In the histopathological differential diagnoses, sarcomatoid carcinoma, endometrial stromal sarcoma and Mullerianadenosarcoma must be considered.[6]Insarcomatoid carcinomas, there is always a sharp merging between the obvious epithelial component and the sarcomatoid component, whereas this merging is not seen to the same degree in MMMTs. The distinction between MMMT and Mullerianadenosarcoma is easier because in adenosarcoma, the epithelial component is clearly benign. Endometrial stromal sarcomas arising in the cervix is extremely rare. Only three cases have been reported so far and the tumor can be differentiated from MMMT by absence of a malignant epithelial component. MMMTs may be misdiagnosed as pure carcinomas or sarcomas, especially in small or inadequate biopsies. Also, because of its rarity, the cervical extension from the uterine corpus must be excluded since this condition is more common and most cervical MMMTs are microscopically indistinguishable from its endometrial counterpart. In such cases, the correct diagnosis depends mainly on the dominant localization of the neoplasm based on the findings of pelvic

examination, imaging studies, curettage and in some patients, a hysterectomy specimen. In our case, cervical origin was confirmed on hysterectomy specimen. Another matter of persistent controversies is the histogenesis of MMMTs of the female genital tract. Theories which have been proposed include the "collision", "combination" and "composition" theories. The fourth and currently favoured theory is the metaplastic theory of histogenesis (Agale *et al.,* 2009). Thistheory is supported by the detection of HPV 16 and 18 in cases of MMMTs of the female genital tract. In a study done by Grayson et al., HPV 16-DNA was detected in the nuclei of both the epithelial and sarcomatous components of three cases. Yet another theory, the "neometaplasia of Mullerian origins", states that mesodermal stem cells differentiate along many divergent cells lines leading to the development of the different elements in MMMTs (Agale *et al.,* 2009). The prognosis is cervical MMMTs depends on the clinical stage of the disease and presence of metastasis (Munakata *et al.,* 2013). Spread ofcarcinosarcomas is primarily via the lymphatic system. The most frequent areas of spread are the pelvis, lymph nodes, lungs and liver (Kuyumcuoğlu and Kale, 2009). Compared to its uterine counterpart, cervical MMMT is more often confined to the uterus at presentation, frequently has non-glandular epithelial component and may have better prognosis. Due to rarity of this tumor, no evidence based management guidelines are available (Agale *et al.,* 2009). Surgery (Radical Hysterectomy) is the principal modality of treatment (Thomas Stovall, 2012). Although adjuvant chemotherapy and/or radiotherapy have been used, their role is not well-defined, in terms of overall survival benefit. Radical radiotherapy with or without chemotherapy is recommended for locally advanced disease. Patients with metastatic disease are treated with palliative chemotherapy. Taxanes and cisplatin based chemotherapy, ifosfamid, along with pelvic irradiation may lead to increased survival in patients with metastatic carcinosarcomas (D'Angelo, 2011). Overall, 5year survival is 20-30%.

Conclusion

Cervical malignant mixed Mullerian tumors are rare, highly aggressive, rapidly progressing neoplasm associated with a poor prognosis. It may at times present diagnostic difficulties to the clinician. They are best treated by surgery with or without adjuvant radiation and/or chemotherapy. But the optimal management modality remains controversial, with discrepancies regarding patient outcome to lymphadenectomy and radiation therapy.

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