Malignant mixed mullerian tumour of uterine cervix with heterologous mesenchymal component– A rare case report

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ABSTRACT

Malignant Mixed Mullerain Tumors (MMMT) of the uterus and cervix are biphasic tumors, commonly composed of non glandular carcinomas and the homologous sarcomatous components occurring in elderly age. These tumours are more common in the body of the uterus and extremely rare in the cervix. We present a case of MMMT of the cervix having endometrioid adenocarcinoma (glandular carcinomatous element) and grade I chondrosarcoma as heterologus mesenchymal component. The tumour had spread to the adjacent areas; however, the body of the uterus was uninvolved. This is a rare event, furthermore, this case is presented to emphasize that this entity has to be considered as a differential diagnosis for the cervical tumors.

KEYWORDS: Malignant Mixed Mullerain Tumors, cervix.

INTRODUCTION

Primary malignant mixed mullerian tumors of cervix are uncommon neoplasms with fewer than 50 documented cases in literature [1]. Malignant mixed mullerian tumors are rare biphasic malignant neoplasm consisting of both epithelial and mesenchymal components. The commonest site of occurrence of MMMT in the female genital tract is uterine corpus. It was first described by Ferriera in 1951 [2]. The most common carcinomatous component is basaloïd squamous carcinoma and the sarcomatous element is usually homologous high grade [1]. We report a case of malignant mixed mullerian tumor of uterine cervix in a post menopausal woman. The carcinomatous component being, endometrioid adenocarcinoma and heterologous sarcomatous element being grade-I chondrosarcoma; which is a rare event.

CASE REPORT

A 55 year old post menopausal woman presented with mass in lower abdomen of one month duration. CT scan showed large irregular heterogeneous soft tissue mass arising from cervix involving the pelvis (Figure 1). Later the patient underwent total abdominal hysterectomy. The specimen was sent as multiple irregular grey white to grey brown friable tissue bits. A small atrophic uterus measuring 4.5 x 3.0 x 2.0cm was found among the friable tissue bits. Both ovaries and tubes were unremarkable.

Hematoxylin and Eosin stained sections from the tumor tissue showed biphasic pattern composed of malignant epithelial and sarcomatous components. The epithelial component was predominantly an adenocarcinoma which showed glands lined by pleomorphic epithelial cells (Figure 2), with some areas of basaloïd squamous cell carcinoma. The sarcomatous component consisted of grade-I chondrosarcoma (Figure 3) which intimately blended with carcinomatous areas. This sarcomatous element was heterologous to the site, which is rare.

The panel of immunohistochemical tests was performed. The epithelial component was positive for cytokeratin (CK) (Figure 4a) but negative for vimentin confirming their epithelial nature. The stromal component was diffusely positive for vimentin and S-100 cytoplasmic stain (Figure 4b).
Figure 1: CT scan showing tumor of cervix extending to pelvis (Arrow)

Figure 2 Microphotograph of adenocarcinoma with glands lined by malignant cells. (H&E, 10X)

Figure 3 Microphotograph of chondrosarcomatous component. (H&E, 40X)

Figure 1 Immunohistochemistry: 4a) Cytokeratin positivity by the epithelial cells. 4b) Sarcomatous component showing vimentin positivity. Inset shows S-100 positivity in the chondromatous areas.

DISCUSSION

The cervical MMMT account for < 3% of all uterine MMMTs[1]. The MMMT occur predominantly in post menopausal women with mean age of presentation being 65-69 years [1]. However the age range in the literature varies from 12 to 93 years. The most common clinical presentation in cervical MMMT is vaginal bleeding or spotting or polypoidal cervical mass and less commonly an abnormal pap smear [1, 2]. However, our case presented with lower abdominal mass which is similar to a case report by Maheshwari A et al[2] and Kadota K et al[3].

Gross examination showed predominantly polypoid large grey white mass in most of the studies, however our case had friable multiple gray white bits and the body of uterus was amongst these bits which was unremarkable. According to the earlier case reports [2, 4] the epithelial component comprised mainly the basaloid squamous cell carcinoma. Adenocarcinoma as the predominant epithelial component is rarely described in literature. Agale VS et al [1] described the presence of adenocarcinoma as a predominant epithelial component in their cases which is similar to our case. Studies have suggested that presence of non-glandular epithelial component have an impact on the clinical behavior of such cases.

The sarcomatous component may be homologous (fibroblasts and smooth muscle) or heterologous (cartilage, striated muscles, bone etc) [2]. In literature homologous sarcomatous component is more frequent than the heterologous component. Our case has the heterologous chondrosarcoma as its sarcomatous component. However study by Agale VS et al [1] showed rhabdomyosarcoma and malignant spindle cells mixed with mature cartilage as their heterologous sarcomatous component. Kadota K et al [3] published a case report which had endometrioid adenocarcinoma and chondrosarcoma as its biphasic components, which is similar to our case.
The immunohistochemical studies revealed that CK was positive in epithelial component, while vimentin, desmin, SMA were positive in the sarcomatous component [2]. However in our case CK was positive in epithelial component and S-100 and vimentin was positive for the sarcomatous component which establishes the diagnosis of chondrosarcoma.

The histogenesis of MMTT of the female genital tract remains a long standing enigma and controversies. Grayson et al [4] studied human papilloma virus (HPV) status in eight patients with cervical MMTT. In all cases HPV-DNA was detected by polymerase chain reaction. By the in situ hybridisation technique HPV-16 DNA was demonstrated in the nuclei of both epithelial and sarcomatous components in their cases. This observation supported the metaplastic theory of histogenesis. The immunohistochemical marker P53 was positive in the epithelial and mesenchymal elements in a case study by Koda K et al [3]. This may be one of the finding explaining histogenesis by combination theory.

Due to rarity of this tumor, no evidence based management guidelines are available. Surgery is the principal modality of treatment, although adjuvant chemotherapy and or radiotherapy have been used. Their role is not well defined. Radical radiotherapy with or without chemotherapy is recommended for locally advanced disease. Patients with metastatic disease are treated with palliative chemotherapy [2].

The clinical behavior of these tumors is dominated by the carcinomatous component. It has been suggested in literature that long term survival is possible in organ confined early stage disease with primary therapy. Our patient was in an advanced stage at presentation and was treated with surgery, but her condition was poor after treatment.

CONCLUSION

To conclude, cervical MMTT is an unusual cervical malignancy and must be considered in the differential diagnosis of cervical tumors, in post menopausal women presenting with vaginal bleeding or abdominal mass. Since most cervical MMTT’s are confined to uterus and have better prognosis when treated by surgery with / without adjuvant therapy; their detection is critical for appropriate treatment and prognostication of the disease.

REFERENCES


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