International Journal of Clinical Obstetrics and Gynaecology

ISSN (P): 2522-6614
ISSN (E): 2522-6622

www.gynaecologyjournal.com
2018; 2(3): 37-39
Received: 01-03-2018
Accepted: 04-04-2018

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A rare case report of vault carcinoma of malignant mixed mullerian tumor in a post hysterectomy patient

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Abstract
Introduction: Carcinosarcoma is composed histologically of a mixture of sarcoma and carcinoma. Malignant mixed mullerian tumors (MMMT) are rare biphasic malignant neoplasm. The commonest site of occurrence of female genital tract MMMT is the uterine corpus. Predominantly these carcinomas are found in postmenopausal women presenting with uterine bleeding and uterine enlargement.
Case Summary: A 52 years P4L4 female presented with Postmenopausal bleeding and abdominal pain. Patient underwent hysterectomy 10 years back for menorrhagia. On Per speculum examination a 8cm X 5cm firm to hard and rubbery growth was seen involving the anterior vaginal wall. On per vaginal examination growth was found to be involving the bilateral fornices and it bleeds on touch. On Ultrasound findings of a large well marginated rounded and solid hypoechoic mass with internal septations in the vault region. Mass measuring approximately 8.4 X 7 cm? Cervical mass. She underwent Exploratory laparotomy followed by removal of vault mass and left salpingoophorectomy. A mass of around 10 x 7 x 7 cm globular mass involving anterior vaginal wall was removed. It was indenting the posterior wall of bladder. On HPR, the features were suggestive of Poorly Differentiated Malignant Mixed Mullerian Tumor.
Conclusion: Malignant mixed Mullerian tumors are extremely rare and aggressive tumors. There should be a high index of suspicion for a postmenopausal hysterectomised patient presenting with per vaginal bleeding and should consider the possibility of malignant mixed Mullerian tumor. The outcome depends on the stage of disease and the depth of invasion. Combination chemotherapy and radical surgery remains the mainstay of treatment

Keywords: MMMT malignant mixed mullerian tumors

Introduction

Primarily Vaginal Carcinoma contributes to only 2-3% malignant neoplasms of female genital tract. However, Majority of cancers found in the vagina arise from cancers metastatic to vagina. The FIGO staging of Vaginal Cancer dictates that a tumor extending to the vagina from the cervix be regarded as a cancer of the cervix, whereas a tumor involving both the vulva and the vagina should be classified as cancer of the vulva. Staging is performed by clinical examination and, if indicated, cystoscopy, proctoscopy, and chest and skeletal radiography. Surgical staging and resection of enlarged lymph nodes may be indicated in selected patients.

Figo Staging Of Vaginal Cancer	
Stage i	The carcinoma is limited to the vaginal wall
Stage ii	The carcinoma has involved the subvaginal tissue but has not extended to the pelvic wall
Stage iii	The carcinoma has extended to the pelvic wall
Stage iv	The carcinoma has extended beyond the true pelvis or has involved the mucosa of the bladder or rectum; Bullous edema as such does not permit a case to be allotted to stage IV IV A - Tumor invades bladder and/or rectal mucosa and/or direct extension beyond the true pelvis IV B - Spread to distant organs

The association of cervical cancer with HPV suggests that vaginal cancer may have a similar association. By convention, any new vaginal carcinoma developing atleast 5 years after cervical cancer is considered a new primary lesion. Cancer of the vagina spreads most often by direct extension into pelvic soft tissues and adjacent organs. Metastases to the pelvic and para- aortic lymph nodes may occur in advanced disease. Lesions in the lower one-third of the vagina may spread directly to inguinal femoral lymph nodes and the pelvic nodes. Hematogenous dissemination to the lungs, liver or bone may occur as a late phenomenon.

Most common form of vaginal cancer is Squamous cell carcinoma occurring in 80 to 90% of cases. Malignant melanoma is second most common cancer of the vagina, accounting for 2.8% to 5% of all vaginal neoplasms. Primary adenocarcinomas of the vagina is rare, constituting 9% of primary tumors of the vagina. The most common adenocarcinoma of the vagina is metastatic, originating from the colon, endometrium, ovary, or rarely pancreas and stomach. However, most common malignant mesenchymal tumors of the vagina in adult women and children and infants Smooth muscle tumors and Botryoid Rhabdomyosarcoma.

Carcinosarcoma is composed histologically of a mixture of sarcoma and carcinoma. Almost all of these tumors occur after menopause at a median age of 62 years. The most frequent presenting symptom is postmenopausal bleeding which occurs in 80% to 90% of cases. The most important single factor affecting prognosis in patients with carcinosarcoma is the extent of tumor at the time of treatment.

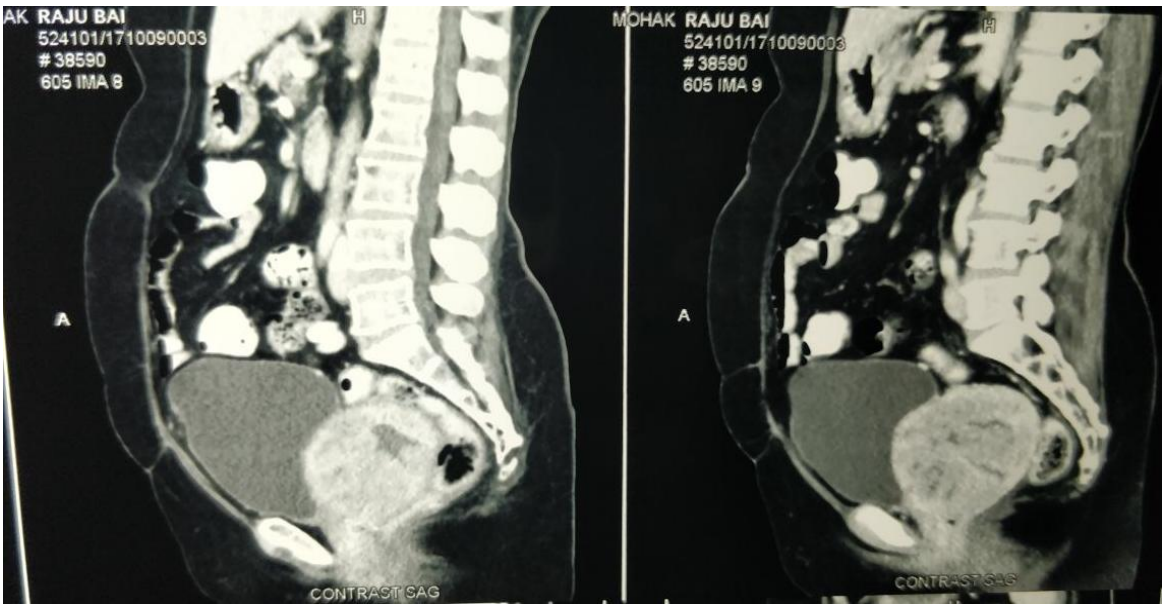
Classification of Uterine Sarcoma

Pure Nonepithelial Tumor		**Mixed Epithelial-Nonepithelial Tumor**
Homologous	**Heterologous**	Malignant Mixed Mullerian Tumor
Endometrial stromal tumors	Rhabdomyosarcoma	
Smooth muscle tumors	Chondrosarcoma	Adenosarcoma
	Osteosarcoma	
	Liposarcoma	

Case summary: A 52 years P4L4 female presented to MYH, Indore with Postmenopausal bleeding with chief complaints of Bleeding Per vaginum since 15 days,Heavy bleeding since 4-5 days. Patient underwent hysterectomy 10 years back for menorrhagia. On Per speculum examination a 8cm X 5cm firm to hard and rubbery growth was seen involving the anterior vaginal wall. On per vaginal examination growth was found to be involving the bilateral fornices and it bleeds on touch. Patient got herself investigated in some private hospital and had findings of a large well marginated rounded and solid hypoechoic mass with internal septations in the vault region. Mass measuring approximately 8.4 X 7 cm? Cervical mass.

On Biopsy from anterior vaginal wall reaveal vaginal mucosa lining stratified squamous epithelium showing hyperplasia and mild koilocytic changes. Subepithelial tissue show interstitial hemorrhage and congested blood vessels. No evidence of malignancy or dysplasia in section studied.

CT Pelvis Report: It shows a lobulated heterogeneously enhancing mass lesion of approx. 10 x 7.5 x 7.4 cm (CCxMLxAP) in size situated posterior to urinary bladder and anterior to rectum. The mass is elevating the posterior wall of urinary bladder anteriorly and there is loss of fat plane between the mass and urinary bladder. The mass is indenting rectosigmoid posteriorly. The mass is seen in contiguity with the vagina. Pelvic side walls appear free. Findings are suggestive of mitotic etiology. Rest of the findings were within normal limits.



CT scan pelvis: Sagittal view showing the presence of a huge mass in the pelvic cavity

Intraoperative Findings: A mass of around 10 x 7 x 7 cm globular mass involving anterior vaginal wall was removed. It was indenting the posterior wall of bladder. She underwent Exploratory laparotomy followed by removal of vault mass and left salpingoophorectomy.

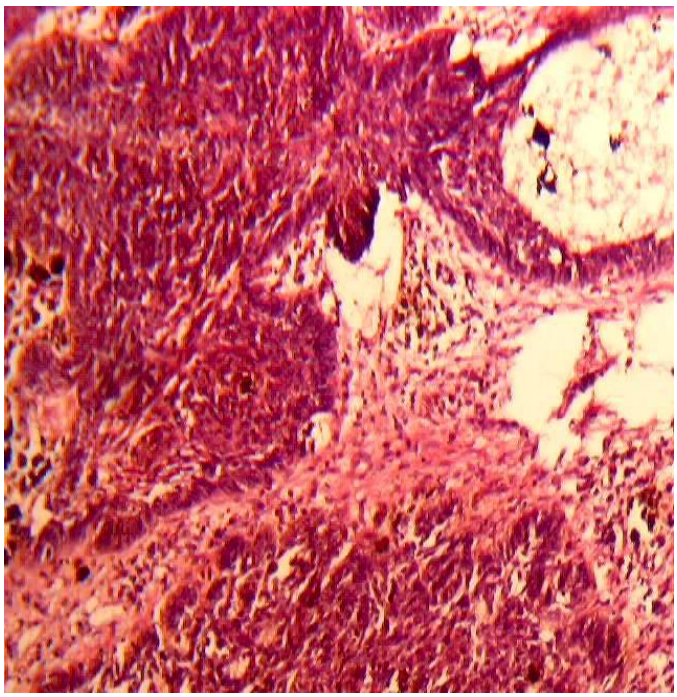
Intraoperative specimen of growth obtained from the anterior vaginal wall.

HPR Report: On her HPR report in the section studied show biphasic tumor with markedly pleomorphic tumor cells arranged in the form of glands and spindle cells arranged in form of bundle and fascicle. Mitotic index is raised.

- Histological Type: Carcinosarcoma (Malignant Mixed Mullerian Tumor)
- Histological Grade: Poorly differentiated (Grade III)
- Degree of invasion: Upto vaginal vault
- Extent of tumor: Upto vaginal vault
- Lymphvascular invasion: Present
- Lymph node status and status of resection margins: Could not be assessed
- Tumor marker status: Not known
- Adjoining pathology: Non specific inflammation with areas of hemorrhage and necrosis

Comment: Features suggestive of Poorly Differentiated Malignant Mixed Mullerian Tumor

Diagnosis: Malignant Mixed Mullerian Tumor



10x view showing biphasic pattern of growth comprising of islands of epithelial cells and sarcomatous component in the form of sheets of spindle shaped cells

Discussion

In the preceding studies it was found that most of the Vaginal Vault cancers were found in association with the residual cervical cancer or in cases of subtotal hysterectomy with cervical stump in situ. It was mostly metastatic with very less primary arising from the vault itself. Vaginal Vault cancer was found as a second primary in a treated case of ovarian cancer. In my Case report it was found to be Malignant Mixed Mullerian Tumor which is a type of carcinosarcoma. This is one of the rare variety seen in association with vault carcinoma in a post hysterectomy patient.

References

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