IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 21, Issue 1 Ser.12 (January. 2022), PP 50-52
www.iosrjournals.org

The Unusual Triad of a Rare Uterine Malignancy-Carcinosarcoma

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Abstract
Carcinosarcoma is a rare type of uterine malignancy comprising of less than 5 % of the uterine malignancy. They are also known as mixed malignant mullerian tumors. They are undifferentiated tumours with carcinoma and sarcomatous component, arising from a single malignant epithelial cell. The clinical presentation is distinct and non specific imaging and pathology studies can lead to a late diagnosis and treatment. This report deals with a peculiar presentation of a case of uterine malignancy and the further management planned.

Date of Submission: 07-01-2022 Date of Acceptance: 21-01-2022

I. Case Report

A 55yr thin built P2L2 post menopausal since 6 yr came to the opd with c/o pain in lower abdomen and mass protruding out from the vagina with minimal white discharge per vaginum since 3-4months. General examination was normal with no significant medical history. On proceeding with the per speculum and pervaginal examination a large fleshy mass was seen protruding from the utrine cavity with the cervix taken up and forming a ring around the mass no evidence of discharge or bleeding was seen. There was no ulceration or erosion but slight areas of necrosis were seen. Pervaginal examination confirmed the finding and estimated the uterine size to be around 10-12 weeks with a large mass felt with a thin rim of cervix that was free. The pedicle of the protruding mass could not be felt. The mass was thought to be a polyp. Some tissue from the mass sloughed out and was sent for histopathological examination.

Meanwhile the patient was investigated further and use done revealed a large mass completely occupying the whole uterine cavity, no pedicle present, enlarged uterus and ET was increased. Patient was planned for D & C with polypectomy following which the day after she c/o discharge from the vagina. However this time the examination left us astonished and the mass that was seen earlier could not be felt . There was serous discharge and a pap and vaginal swab was taken and sent. But the per vaginal examination sill revealed an enlarged uterus. However this time the repeat USG, did not demonstrate any growth. Hence, we estimated that the mass would have become necrosed and fallen off and patient was treated for the vaginal discharge as she also c/o post menopausal bleeding, patient was evaluated and posted for fractional curettage.

Curettage sample revealed fleshy and cheesy with material with profuse endometrium. The histopathological report showed that we were dealing with a case of uterine carcinoma of a rare variety ie carcinosarcoma. The patient was planned for CT scan and then posted for exploratory laparotomy. CT was

s/o uterine size of 8.4*6.2*10.2 with heterogeneous enhancement with myometrial thinning, mild hydronephrosis but no bladder involvement with few para aortic , pre caval and subcentrimetric enlarged lymph nodes of 6mm approx. after the exploratory laparotomy patient received chemoradiotherapy. Her follow up CT scans done after the cycles were s/o no lymphadenopathy or any other organ involvement.

II. Discussion

Uterine carcinosarcomas are highly aggressive, rare biphasic tumours composed of epithelial and mesenchymal components believed to be of monoclonal origin(1). The name malignant mixed mullein tumour originates from the observation that the mullein or the paramesonephric ducts created from the intermediate mesoderm of the coelomic epithelium invaginate lateral to the mesonephric duct at the 6th week of embryogenesis and form the female genital tract ie the fallopian tube, uterus, cervix and cranial portion of vagina. In males mullerian tube regress due to the AMH derived from the sertoli cells. These tumors arise in the uterus and also in vagina cervix and ovaries in decreasing order and rarely from female peritoneum(2). Looking

in the back dates the various names given to this are enchondroma, malignant mesodermal mixed tumors, metaplastic carcinoma and carcinosarcoma.

These represent less than 5% of the uterine cancers and account for 16.4% of all deaths. Risk factors include nulliparity, advanced age, obesity, exposure to exogenous estrogens, long term use of tamoxifen and also radiation exposure. Tamoxifen use is associated with 2-7 times greater risk of development.(2)

Carcinosarcomas have two histological types- the sarcomatous component and the epithelial component. The sarcomatous component can be homologous or heterologous(1). The heterologous types are rhabdomyosarcoma, chondrosarcoma or liposarcoma while the homologous components include fibrosarcoma, endometrial stroll sarcoma or leiomyosarcoma. The theories(1) proposed for its origin are:

A) The collision theory- stating that the two components had separate point of origin prior to colliding together to form a single tumour.

B) The combination theory- a common stem cell precursor undergoes bidirectional differentiation that results in the creation of two histological types

C) Conversion theory- hypothesises that a single epithelial component undergoes metaplastic differentiation from which the mesenchymal components arise.

It is believed that thee have a single monoclonal origin from a common progenitor stem cell.

The epithelial component is the driving component.

Clinical features-

A typical presentation includes pyometra with vaginal bleeding, watery or bloody discharge,abdominl pain or a polypoid mass protruding from the endocervical canal.

Pathology

Grossly they appear as solitary polypoidal mass with areas of haemorrhage and necrosis projecting from the uterine cavity.gritty or hardened areas suggest osseous or cartilaginous differentiation. The carcinomatous component arise from the posterior wall of uterus , fills and distends the whole uterine cavity. Tumours are bulkier and fleshier than the endometrial adenocarcinoma(3).

Microscopically the two components may be mixed or seen distinctly. The epithelial component is often high grade papillary serous(66%),or endometriosis type (42%). Solid areas of marked pleomorphism with bizarre cells embryonal glandular components may be present.

Carcinosarcoma express epithelial membrane antigen (EMA), pancytokeratin and stroll lineage markers like design or S100. Over expression of HER-2 , EGFR and KIT are seen. Commonly associated mutations include TP53 ,PIK3K, PTEN,KRAS, PPP2R1A, CHD4 and BCOR.

DIAGNOSTIC IMAGING

Early cancers in ultrasound present as hyperechoeic mass with a thickened endometrial stripe with expansion of the endometrial canal(4).

On comparing with other endometrial cancers heterogeneous mass with a greater craniocaudal width is suggestive of carcinosarcoma. On MRI enhanced uterine lesion with enhancement equal to greater than the myometrium suggests this tumour type.

CT scans can easily confuse this with leiomyosarcoma or endometrial carcinoma.

F-PDG PET scan can show metastasis . The principal behind this is that malignant tissue show greater glucose metabolism than benign.however due to the cost this is not very widely reported.

TREATMENT

The primary treatment remains surgery with post operative adjuvant therapy in the form radiotherapy/chemotherapy is essential due to high rate of relapse and metastasis.

This multimodality treatment of surgery combined with postoperative radiotherapy/chemotherapy increases the disease free survival

(DSS- 31months) as compared with surgery alone(DSS- 3months) (4).

Surgery -

As we struggled with the fact weather to remove the lymph nodes or not the literature proposes a controversial role of lymphadenectomy(2).

The role of pelvic and paraaortic lymphadenectomy, technique and the optimal number of lymph nodes to be removed remains undetermined(2). Three primary arguments have been put forward regarding this:

A) accurate staging with lymphadenectomy can allow for determination of the patients true metastatic potential.

B) Possible reduction in the locoregional recurrence.
C) Improving the selection of patients for adjuvant therapy.

However ,lymphadenectomy provides a survival advantage only in node negative patients, as removal of lymph nodes in positive patients can upstage the disease and worsen the prognosis.
But negative nodes can also have micrometastasis that if removed can improve survival.
A recent publication by Garg et all concludes that optimal patient management includes abdominal hysterectomy with b/l salpingo-oophorectomy, lymph node dissection, resection of any gross abdominal disease and sampling of peritoneal washings.

Radiotherapy

Radiation therapy in the form of whole abdomen radiotherapy or vaginal brachytherapy and offers benefits in preventing the loco regional recurrence or intraperitoneal seedings.
This is because few patients are upstaged by surgical staging compared with clinical staging.

Chemotherapy

Extra pelvic recurrence /relapse occur due to hematogenous, lymphatic and transcoelomic spread. Chemotherapy has a role in minimising these.
Active single anti neoplastic agents include ifosfamide, cisplatin, doxorubicin and paclitaxel.
Combination of cisplatin with ifosfamide has shown highly effective.
However treatment with these has more toxic effects on treatment in comparison to uterine carcinoma.
These agents target the biological agents like all, Her-2 , PDGFRbeta etc.
Elevated Ca-125 levels postoperatively confers increased risk of death.

RECURRENCE

Mostly occurs within one year. Even in early disease the rate of recurrence is 47-64%(2). Factors like patients age, extent of myometrial invasion, tumour size, lymphovascular space involvement, histological grade, peritoneal cytology and cell type are important determinate of recurrence and distant metastasis.metastatic disease are usually asymptomatic and can spread by both lymphatic and vascular route as compared with endometrial cancers.the most common sites (3)of metastatic foci include- lungs(49%), peritoneum(44%), pelvic or paraaortic nodes(35%), adrenal glands, pericardium brain etc. uterine carcinosarcomas have the highest rate of pulmonary metastasis as compared with other uterine malignancy(3).

PROGNOSIS-

Although rare these account for about 15 % of the uterine cancer related deaths. Stage is an independent prognostic variable for predicting survival. stage1/2 have overall 5 year survival rate of 30-46% while stage 3/4 have only 0-10%.(5).
Characteristics like myometrial invasion less than 1/3rd of myometrium, size less than 7cm, no detectable distant metastasis are associated with favourable prognosis. Also the tumour histology plays a role with homologous type having a favourable prognosis than heterologous type. Positive peritoneal cytology and tumour markers like p53 in older women have shorter median survival time.

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Dr Sangeeta Ramteke, et. al. "The Unusual Triad of a Rare Uterine Malignancy-Carcinosarcoma." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 21(01), 2022, pp. 50-52.