A Case Report on Rhabdomyosarcoma of the Uterus

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Abstract:
Most uterine sarcomas fall into these categories: leiomyosarcoma, endometrial stromal sarcoma or undifferentiated sarcoma. Yet rhabdomyosarcoma element may be present as a component of adenosarcoma or carcinosarcoma (malignant mixed Mullerian tumor). Pure rhabdomyosarcoma are extremely rare. We report a case of a 60 year old postmenopausal woman para 6 live 6 came with complaints of brownish discharge per vaginum. Ultrasoundography showed bulky uterus with pyometra. Fractional curettage revealed malignant mixed Mullerian tumor. Patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy with pelvic lymphadenectomy. Rhabdomyosarcoma with review of literature is presented in the following work.

Key word: HETEROLOGOUS Rhabdomyosarcoma Uterus

REVIEW OF LITERATURE:
Anderson and Edmansson reported the first rhabdomyosarcoma of the corpus uteri in 1869, and the first English report was by Robertson in 1909. Donkers and colleagues in 1972 reported two cases and reviewed the literature. They were able to find 49 rhabdomyosarcomas of the corpus uteri reported since 1869. Except for reports by Kukla and Douglas and Middlebrook and Tennant, most studies are reports of one to three patients. A 33-year review of all uterine sarcomas at the University of Iowa revealed no pure corpus uteri rhabdomyosarcomas. We find that the rarity of this histological entity makes it particularly worthy of publication.

CASE REPORT:
60 year old postmenopausal woman para 6 live 6 came with complaints of non-foul smelling brownish discharge per vagina for 1 month associated with lower abdominal pain and abdominal distention for 10 days. On examination general condition was fair and vitals were normal, systemic examination was normal. Per abdomen examination, abdomen was soft and no organomegaly. Per speculum examination revealed pin hole OS with drainage of blood stained fluid. Per vaginal examination uterus was progressively increasing in size around 12 to 14 weeks due to collection of fluid and growth of mass. On investigating Pap-smeared showed high grade squamous intra epithelial lesion. Ultrasound abdomen showed bulky uterus with heterogeneous echoes and irregular anechoic areas (7 x 4.8 x 2.8) sized irregular collection in the body of uterus. Cervix appears normal. Ovaries not visualized separately. Fractional curettage done and histopathology revealed malignant mixed Mullerian tumor with heterologus rhabdomyosarcomatous component. CT Pelvis showed Grossly enlarged uterus (9x12x10.5) with distended endometrial cavity with irregular enhancing lesions and large amount of fluid.
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FIG3&4: SPECIMEN OF UTERUS & ITS CUT SECTION SHOWING MALIGNANT GROWTH
Total Abdominal Hysterectomy with Bilateral Salpingo Oophorectomy with pelvic Lymphadenectomy done. Histopathology showed Malignant mixed mullerian tumor with heterologous (rhabdomyosarcomatous elements) Invasion into inner half of myometrium Both Ovaries and Fallopian tubes are free of tumor. lymph nodes were free of tumor

FIG5&6: TUMOUR COMPOSED OF PLEOMORPHIC SPINDLE SHAPED CELL WITH ABUNDANT EOSINOPHILIC CYTOPLASAM

FIG7: IMMUNOHISTOCHEMISTRY POSITIVE FOR CYTOKERATIN

FIG8: IMMUNOHISTOCHEMISTRY POSITIVE FOR MYOGENIN
Post operative period was uneventfull.

DISCUSSION:
Malignant mixed mullerian tumor and carcinosarcoma, is a malignant neoplasm found in the uterus, ovaries, fallopian tube and other parts of the body that contains both carcinomatous (epithelial tissue) and sarcomatous (connective tissue) components. It is divided into 2 types. Homologous in which the sarcomatous component is made of tissues found in the uterus such as endometrial, fibrous and or smooth muscle tissues. Heterologous type (made of tissues not found in the uterus, such as cartilage, skeletal muscle & or bone) Malignant mixed mullerian tumor account for between 2-5% of all tumors derived from the body of uterus and are found predominantly in postmenopausal women with an average age of 66 yrs. Risk factors are similar to those of adenocarcinoma & include obesity, exogenous estrogen therapies & nulliparity. pure rhambdomyosarcoma of uterine corpus can be encountered in 2 distinct histopathological contexts usually as a component of malignant mixed mullerian tumour & exceptionally as a pure heterologous sarcoma. Immunohistochemical analysis is necessary to distinguish between these two entities. Immunohistochemical stain positive for CYTOKERATIN & MYOGENIN. only the absence of epithelial components, after extensive tumour sampling, can exclude malignant mixed mullerian tumour. Tumour primarily metastasize – locally in pelvis & lungs.

PROGNOSIS:
Outcome of mixed malignant mullerian tumour is determined primarily by depth of invasion & stage, pelvic lymphnodes may show metastases in 17% cases where tumor grossly appears confined to uterus. In Early stage disease 5 year survival rate 40 -50 %. In Advanced disease 5 year survival rate is 25 – 30%. Recurrence develop in more than one half of cases of uterine sarcoma, even when disease is apparently localized at the time of treatment. Based on evidence, stage 1 & 2 total abdominal hysterectomy with bilateral salpingo oophorectomy and treatment of pelvic lymphatic’s by irradiation or surgery.

REFERENCES: