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REVIEW ARTICLE

Rare case of cervical leiomyosarcoma

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ABSTRACT

The most common histological subtype of cervical cancer is squamous cell Carcinoma, which is followed by adenocarcinoma, adenosquamous and small neuroendocrine carcinoma. Cervical sarcomas are rare tumors that constitute less than 1% of all cervical malignancy. Due to rarity of this tumor it is difficult to have a randomized study to guide treatment and analyze prognostic factors with the result treatment is not standardized.We present a rare case of cervical leiomyosarcoma and review of literature.

INTRODUCTION

Case Summary

A 48 years old married lady and with two healthy children, not a known diabetic or hypertensive. She has undergone thyroidectomy 16 years back. for benign pathology. She had presented in 2014 to her local hospital with symptoms of menorrhagia and polymenorrhea. She was investigated and found to have a large fibroid and was treated with abdominal hysterectomy and bilateral salphingo-ophrectomy. Histopathology was reported as low grade leiomyosarcoma arising from cervix with normal uterine cavity. She was given adjuvant chemotherapy of gemcitabine and docetaxel (5 cycles). In 2015, she again complained of bleeding per vaginum and on investigation was found to have central pelvic recurrence involving the vaginal vault.

Repeat biopsy from vaginal vault was reported as low grade Leiomyosarcoma. Her staging work up at that time was negative for metastasis. She received 3 more cycle of Adriamycin with progression of disease, clinically and radiologically. She presented to our hospital in March 2016 with symptoms of pain and lump in lower abdomen and bleeding per vaginum. On questioning further, she gave history of occasional fever with chills and constipation though she refused any history of having hematuria or bleeding per rectum. Physical examination showed an infraumblical midline scar with two palpable nodules on left side of scar each measuring approximately 3 X 3 cm. A 20X20 lump was palpable in lower abdomen arising from pelvis, which was firm in consistency, non-tender with lower margin not reachable. On per vaginum examination, there was a large proliferative growth involving the vault, friable in nature, while rectal examination showed indenting of anterior rectal wall at approximately 6 cm from anal verge. The rectal mucosa was easily mobile on the lump. FDG PET-CT scan revealed a large FDG avid heterogeneously enhancing well marginated soft tissue mass arising from pelvis. The fat planes with posterior wall of urinary bladder were indistinct compressing both ureters with bilateral hydrouretero-ne phrosis. There were FDG avid abdominal wall deposits on either side of midline in infraumblical region.There were no liver / lung metastasis reported (fig 1, 2, 3).

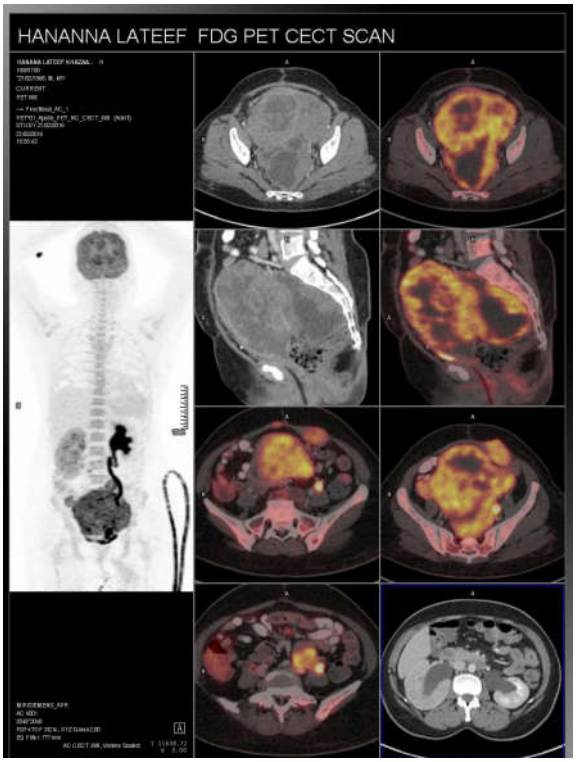

Figure 1, 2 & 3 the collage has a MIP image, axial section and sagittal section of the lobulated pelvic mass lesion (both CT and PET CT), axial images of abdominal muscle deposits (PET CT images), axial section of left common iliac node (PET CT) and axial Image of bilateral hydronephrosis (CT image)

Patient was counseled along with her husband and option of pelvic exenteration with urinary diversion was explained to them. She received pre- operative antibiotics in view of urinary infection. On exploratory laparotomy a large abdomino-pelvic massarising from centre of pelvis with sigmoid colon attached on its superior aspect and urinary bladder stretched on it inferior surface. Both lateral pelvic walls were free. Supralevator pelvic exenteration was done. Bowel continuity was restored by performing Stappled Colorectal anastomosis. Urinary reconstruction was done by ileal conduit. Diversion ileostomy was made. Post-surgery recovery of patient was uneventful. At review at 6 months, she is free of disease, has good anal tone and normal renal functions.

Histopathological Examination

Gross Examination showed a large tumor measuring 17.5x17x13.5 cms. The tumor is composed of long diffuse sheets interwoven fascicles the fascicles are formed by spindle cells showing frequent mitosis; moderate deeply eosinophilic tapered cytoplasm and large ovoid to blunt ended fusiform nuclei with hyperchromasia and pleomorphism. All resection margins are free of tumor (fig 4, 5, 6). The tumor cells show positivity for SMA and negative for CD34, C-kit, DOG -1. Histopathology report is suggestive of high grade Leiomyosarcoma.

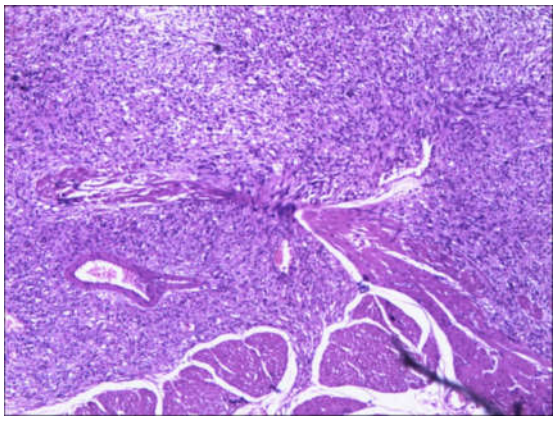

Figure 4 0415- 4X cellular spindle cell tumor with smooth muscle differentiation, interdigitating with normal smooth muscle of Muscular smooth muscle of muscularispropria.

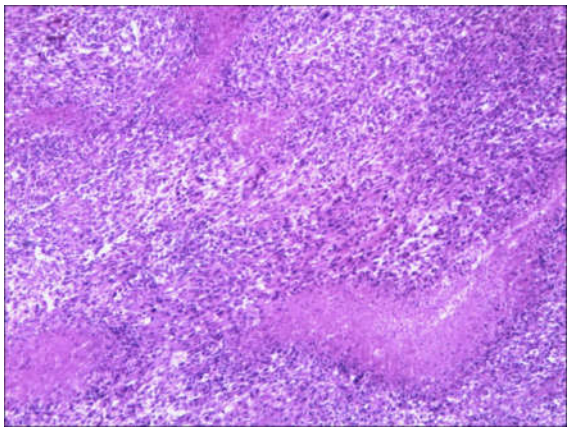

Figure 5 0417 4X large geographic areas of coagulative necrosis seen within the tumor.

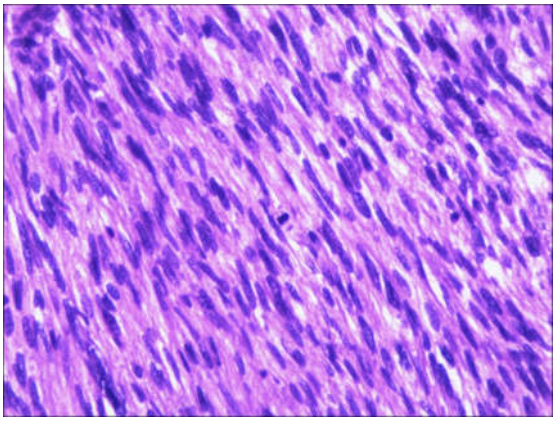

Figure 6 0418 – 20X Atypical smooth muscle cells under higher magnification showing frequent mitoses. Cigar shaped nuclei With moderate pleomorphism and deeply pink cytoplasm.

Discussion

Cervical leiomyosarcoma is an extremely uncommon tumor compared to uterine leiomyosarcoma (LMS). It constitutes less than 1% of all cervical malignancies (1). They generally occur in the premenopausal and post menopausal patient in their 4^{th} -6^{th} decade of life (2). Usual presentation is occasional foul smelling vaginal discharge, bleeding per vagina, pain lower abdomen and bulky cervical mass at the time of diagnosis. Due to rarity of the disease; most of the available data on natural history and treatment of cervical sarcomas are derived from case reports and small case series. Cervical sarcomas consist of several entities that display distinct clinical and pathological profiles. Histopathology may exhibit skeletal or smooth muscle, fatty, fibrohistiocytic, neural sheath, neuroectodermal component and tumors with uncertain differentiation. The most common cervical sarcomas listed in descending order are Embryonalrhabdomyo sarcomas, leiomyosarcoma, undifferentiated endocervical sarcomas and alveolar soft part sarcoma. Ewings and leiomyosarcoma may display spectrum of morphologic subtypes similar to uterine leionyosarcoma including the myxoid variant and epithelialvariant (3). Several prognostic indicators include age. tumor size, stage, grade, mitotic count and menopausal status of the patient. Larger tumors more than 5cm, age > 51 years and postmenopausal status are associated with reduced likelihood of survival in cervical leiomysarcoma (4). Disease confined to uterine cervix in treated with total abdomen hysterectomy with bilateralsalpingo- oophrectomy. Value of routine pelvic lymphadenectomy is questionable. The role of adjuvant chemotherapy is not well defined. Owing to tendency ofhaematogenous spread, most patients are given adjuvant systemic chemotherapy. The current approach is to extrapolate information from the uterine corpus tumors and apply them to cervical counter parts. Usually combination of doxorubicin and ifosfamide are used as first line combination chemotherapy, although chemotherapy regimens are not standardized. The use of adjuvant radiotherapy is debatable due to lack of any large series.Combined use of surgery, radiotherapy and chemotherapy should be used in these patients to achieve better survival (5), complete surgical removal being most paramount.

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