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PRIMARY PAPILLARY ADENOCARCINOMA OF VAGINA: A RARE MALIGNANCY

Oncology

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ABSTRACT

Background-Primary carcinoma of the vagina is rare malignancy of female genital tract which constitutes 1-2% of all gynecological malignancies. The peak incidence is in the sixth and seventh decades of life. Approximately 50% of vaginal cancers arises on the posterior wall of the upper one third of the vagina.

The majority of primary vaginal malignancies are squamous cell carcinoma constitutes 85% and the remaining are adenocarcinoma about 15 %. Histological patterns of adenocarcinoma include clear cell carcinoma, adenosquamous, papillary and undifferentiated. Other types are melanomas, sarcomas and unspecified types.

Case report-We report a case of primary vaginal papillary adenocarcinoma in a 61 years old woman presented with symptoms of persistent and profuse clear vaginal discharge since 7 months, vaginal bleeding since 2 months and pain in lower abdominal region since 1 month and USG showed ill defined soft tissue mass seen in lower one third of vagina.

CT scan suggestive of large ill defined mass approximately 4.8 X 2 .7 cm seen in lower vagina and few subcentrimetric pericaecal and mesenteric lymph nodes seen largest measuring 9 mm in short axis. Immunohistochemistry showed CK20, DUPAN 2 positive and CA 125, CK7, serum CEA negative.

Conclusion- This case reported here for its rare presentation in all gynecological Malignancies.

KEYWORDS

Primary Vaginal Carcinoma, Papillary Adenocarcinoma

INTRODUCTION :

Primary carcinoma of the vagina is a rare malignancy of female genital tract which constitutes 1- 2% of all gynecological malignancies.[1] The peak incidence is in the sixth and seventh decades of life. The majority of primary vaginal malignancies are squamous cell carcinoma which constitutes 85% while the remaining is adenocarcinoma constituting 15 %.[2]

CASE REPORT :

A 61 year old hypertensive post menopausal woman presented in our outpatient department on April 2017 with chief complaints of white discharge from vagina since 7 month, which was watery in nature, associated with painless vaginal bleeding since 2 months which was on and off in nature and pain in lower abdominal region since 1 month.

On examination she was moderately built and nourished, vitals were stabled, pallor was present and generalized lymphadenopathy was absent. On systemic examination cardiovascular system, respiratory system & per abdomen findings were normal. There was no palpable mass or organomegaly.

On local examination no abnormality was found on external genitalia. Per speculum examination revealed an ulceroproliferative growth on the lower one third of vagina on the posterior wall, the ulcer measured approximately 5x4 cm in size covered with white discharge and blood. Cervix os was not visualized due to obscured view by large growth.

On per vaginal examination there was large, irregular proliferative growth about 6x4cm size felt in lower third of posterior vaginal wall. It was firm in consistency. Bleeding was present,cervical os was smooth, both fornices were free. Per rectal examination, rectum mucosa was smooth and free from the lesions. There was no significant personal, menstrual, past or family history.

Biopsy of the lesion was advised with a suspicion of malignancy and sent for histopathological examination that turned out primary papillary adenocarcinoma of vagina.

Microscopic picture showed bits of polypoidal mass entangled in blood clots partially lined by papillary glandular epithelium. The papillary fronds are showing evidence of stratification.nuclear atypia and abnormal mitotic figures. The tumor fragments are surrounded by dense & acute and chronic inflammatory infiltrate, haemorrhage and necrosis.

Taking in to consideration vaginal adenocarcinoma is a rare entity, IHC confirmation was done.

Immunohistochemistry showed CK20, DUPAN 2 positive while CA 125, serum CEA were within normal range.Complete hemogram, chest x ray were normal.

Ultrasonography of abdomen and pelvis showed lower one third of vaginal growth on posterior wall. CECT abdomen and pelvis suggestive of large ill defined mass approximately 4.8 X2.7 cm size seen in lower vagina with loss of fat plane with rectum and urinary bladder. Hepatomegaly with grade I fatty changes and multiple cholelethiasis. Cystoscopy and proctosigmoidoscopy revealed no abnormal findings.

On the basis of clinical examination and investigation, patient was diagnosed as a case of primary papillary adenocarcinoma of vagina. Based on FIGO classification patient was staged as FIGO stage III.

Management: Looking at the size of the lesion patient has been taken for neoadjuvant Chemo therapy (Nabpaclitaxel + cisplatin) planned for three cycles. We have observed that lesion was progressive in spite of given two cycles of neoadjuvant chemotherapy, then patient switched to EBRT with concurrent chemotherapy. AP– PA portal was planned, 2 Gy per fraction, total 60 Gy with weekly injection cisplatin 30 mg2/iv was given. Patient responded well to EBRT, there was 40 % growth regression in first week of treatment. The treatment period was uneventful. We have achieved complete response at the end of fifth week of treatment. She has also received 3 # of (6 Gy per #) HDR brachytherapy. She was on regular followup with no events of residual disease & recurrence clinically.

DISCUSSION:

Carcinoma of the vagina is one of the rarest malignancy comprising 1-2% of all gynecological malignancies.[1] Carcinoma of the vagina is defined as a primary carcinoma arising in the vagina and not involving the external os of the cervix superiorly or vulva inferiorly.

Squamous cell vaginal cancers account for approximately 85% and adenocarcinoma account for approximately 15% of the cases.[2]

Risk factors includes increasing age, atypical cells in the vagina (VAIN), exposure to diethylstilbestrol (DES), multiple sexual partners, early age at first intercourse, smoking, HIV .

Primary papillary adenocarcinoma is quite a rare entity. Commonest presentation of patient is painless vaginal bleeding in 65-80% of cases.[3] But in our case it was watery discharge per vagina followed by painless vaginal bleeding.

The most common adenocarcinoma of the vagina are metastatic, which constitute the majority of vaginal cancers (80%-90%) originating from the colon, endometrium, ovary, or rarely from pancreas and stomach. Hence primary adenocarcinoma of the vagina is diagnosis of exclusion.

Prognostic factors are stage of the disease and type of the lesion. Survival is reduced in patients who are 60 years and above, are symptomatic at the time of diagnosis, have lesions of the middle and lower third of the vagina, or have poorly differentiated tumours.[4,5]

Kucera H & Vavra N4 studied in 434 patients reported that disease is primarily found in elderly as 78% were found to be older than 60 years of age. Younger patients had a 5-year survival of 50%; patients between 61 and 75 years of age, 41.2%; and those 76 years of age or older, 34.3%.

Squamous cell vaginal cancers spread superficially within the vaginal wall, paravaginal tissues and parametria. Distant metastases occur most commonly in lungs and liver. However, adeno carcinomas predominantly have pulmonary metastases and supraclavicular and pelvic node involvement.[6]

EBRT followed by ICRT remains the primary treatment. Several studies have evaluated external beam radiotherapy for vaginal adenocarcinoma. Frank et al.[7] reported 26 patients with primary vaginal adenocarcinoma treated with external beam radiotherapy and brachytherapy.

The combination of external beam radiotherapy and cisplatinum was described by Samant et al [8]

He studied on 12 patients that were treated with concurrent weekly chemoradiotherapy and proved that it is feasible to deliver concurrent weekly Cis-platinum chemotherapy with high dose radiation, leading to excellent local control and an acceptable toxicity profile. In the present case, concurrent cisplatin appeared to sensitize the tumor to radiation since the tumor responded at a lower dose and at a faster rate than expected. Concurrent chemotherapy with irradiation appears to significantly enhance radiation effects on cancer and cycling epithelial cells.

5 year survival rates for women with stage III disease ranges from 25 % to 58% with local failure rates of 30 % to 75 %.[9] Despite treatment with EBRT and brachytherapy ,only 20% to 30 %of patients with stage III & IV disease achieve local control. Pelvic recurrences occur more often than distant recurrences.[10]

CONCLUSION:

To conclude, primary papillary adenocarcinoma is rare, little is known about its etiology and behavior. Early diagnosis with high index of suspicion is extremely important so that effective treatment can be done with less recurrence with good quality of life. This case reported here for its rare presentation in all gynecological malignancies.

Figure1 on per speculam an ulceroproliferative growth

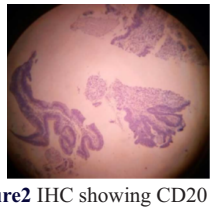

Figure2 IHC showing CD20 positive in 40X

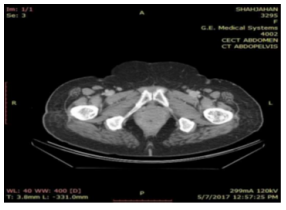

Figure-3 showing lobulated hetrogeniously enhancing mass arising from lower half of vagina, infiltrating into adjacent perineal muscles

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