CASE REPORT

PRIMARY PAPILLARY ADENOCARCINOMA OF VAGINA: A RARE CASE REPORT
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ABSTRACT: Primary vaginal adenocarcinoma is a rare malignant gynaecologic disease. A 56 year-old woman with post hysterectomy status presented with vaginal bleeding. She had a proliferative growth in the posterior vaginal wall extending to the recto-vaginal space and lateral pelvic wall. Vaginal growth biopsy revealed papillary adenocarcinoma and was diagnosed as FIGO stage III primary papillary adenocarcinoma of vagina. As most of the adenocarcinoma of vagina is metastatic, the possibility of primary papillary adenocarcinoma of vagina has to be kept in mind. The present case was successfully treated with chemotherapy and radiotherapy, suggesting that chemoradiation may be an option for the treatment of this type of tumor.

KEYWORDS: Papillary adenocarcinoma, chemotherapy, radiotherapy.

INTRODUCTION: Primary vaginal cancers are rare constitute 1-2% of gynaecological malignancy. Primary adenocarcinoma of the vagina is a rare disease characterized by aggressiveness and poor prognosis because of its rapid growth, recurrence and its frequent distant metastases. We report a case of primary papillary adenocarcinoma of vagina and treated successfully with external beam radiation concurrent with cisplatin followed by brachytherapy.

CASE REPORT: A 56-year-old woman with unconfirmed in utero diethylstilbestrol exposure presented with vaginal bleeding of 3 months duration. She underwent total abdominal hysterectomy with bilateral salpingo-ophorectomy for fibroid uterus 9yrs back. Histo pathology of the uterus, cervix and both ovaries are unremarkable other than leiomyoma. On examination she was moderately built and nourished with BMI 21, vitals were stable. Mild pallor was present and no lymphadenopathy. Abdomen was soft, no organomegaly and no mass felt. Per-speculum examination revealed a proliferative growth of about 2cm x 3cm noted in the posterior vaginal wall in the upper 1/3rd. On rectal examination, a hard mass of about 4cm noted involving the recto vaginal space and parametrium extending up to pelvic wall. Vaginal biopsy revealed papillary adenocarcinoma. MRI abdomen and pelvis shows ill-defined enhancing lesion about 5cm in the posterior vaginal wall involving recto vaginal fat planes and bilateral pelvic lymphadenopathy. Metastatic workup included colonoscopy, upper GI endoscopy, chest X-ray and mammography were normal. Patient was diagnosed with primary papillary adenocarcinoma of vagina FIGO stage III. She was treated with external beam radiation concurrent with cisplatin followed by brachytherapy.

DISCUSSION: Primary vaginal cancer is a rare tumor, representing only 1% to 2% of malignant neoplasms of the female genital tract.¹ Carcinoma of the vagina is defined as a primary carcinoma
CASE REPORT

arising in the vagina and not involving the external os of the cervix superiorly or the vulva inferiorly. Incidence rates for all vaginal cancers combined were 0.18 per 100,000 female population for in situ cases and 0.69 for invasive cases. Squamous cell carcinomas (SCC) are the most common form of vaginal cancer, occurring in 80% to 90% of cases, adenocarcinoma of the vagina is rare, constituting 5-10% of primary tumors of the vagina. The most common adenocarcinoma of the vagina are metastatic, which constitute the majority of vaginal cancers (80% to 90%) originating from the colon, endometrium, ovary, or rarely from pancreas and stomach. Hence primary adenocarcinoma of vagina is diagnosis of exclusion.

SCC and adenocarcinoma of vagina have different natural history of disease course. SCC develops from vaginal intraepithelial lesion following exposure to HPV infection. Many of the reported cases of primary adenocarcinoma of the vagina are clear cell adenocarcinomas in young adolescents exposed to diethylstilbestrol (DES) in utero. The association between the clear cell carcinomas and in utero exposure to DES was first reported in 1971. Vaginal adenosis is most commonly found in young women who had in utero exposure to DES and may coexist or progress to clear cell adenocarcinoma. However, some patients with primary adenocarcinoma of the vagina have non-clear cell, non-DES-associated tumors. The natural history of such tumors has not been clarified.

Glandular elements are occasionally found in the vagina which represents mesonephric or paramesonephric remnants which may undergo malignant change to adenocarcinoma. Adenocarcinomas may arise in foci of endometriosis. Adenocarcinoma of vaginal vault following prolonged unopposed hormone replacement have been reported.

When adjusted for age and prior cervical disease, the incidence of vaginal cancer is not increased in women who have had hysterectomy for benign disease. So women who have had a hysterectomy for benign disease and have no antecedent history of CIN, performance of Pap testing is unnecessary. If the patient has a history of cervical dysplasia or cervical cancer, yearly screening is recommended.

DES associated adenocarcinoma occurs in young girls means age at 19 years, non DES associated adenocarcinoma occurs in older age group between 50-60 yrs. The most common symptom of vaginal carcinoma is abnormal bleeding or discharge. With advanced tumors, pain or urinary frequency occasionally occurs, especially in cases of anterior wall tumors. Constipation or tenesmus has been seen with tumors involving the posterior vaginal wall. These tumors usually are diagnosed by direct biopsy of the tumor mass, and abnormal cytologic findings often will lead to diagnosis of a vaginal cancer.

Non–DES-associated adenocarcinomas generally have a worse prognosis than SCC tumors, and DES-associated clear cell adenocarcinoma.

Therapeutic options depend on tumor stage, age and condition of the patient. Primary therapy for locally invasive disease consists of external and intracavitary or interstitial irradiation, except in selected early cases that may be treated surgically with local excision. Radical surgery, which may involve partial or total pelvic exenteration, is generally reserved for pelvic recurrence after irradiation and for patients with fistulas at diagnosis. Chemoradiation is the newer modality of treatment for primary vaginal cancer. Our patient was treated with external beam radiation concurrent with cisplatin followed by brachytherapy.
CONCLUSION: In summary, primary papillary adenocarcinoma of the vagina is a rare disease associated with a poor prognosis and with significantly worse outcomes than those seen in patients with primary squamous cell carcinoma of the vagina and DES – associated adenocarcinoma. Additional data about patients with this rare tumor should be collected and analyzed in an attempt to elucidate its prognostic factors, characteristics, optimal treatment, and outcome.

REFERENCES:

## CASE REPORT

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