



RESEARCH ARTICLE

PRIMARY MUCINOUS ADENOCARCINOMA OF THE VAGINA: A CASE REPORT

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ABSTRACT

Background and aim: Primary mucinous carcinoma of the vagina is a rare disease which is characterized by aggressiveness and relative resistance to conventional treatment modalities. We aim to report a new case of this rare entity, to investigate its characteristics and its prognostic factors. It is a case of advanced stage of primary mucinous adenocarcinoma of the vagina, diagnosed in our service in January 2015, showing a highly aggressive course.

Case: A 35 year old woman was admitted to our institute to be treated for advanced stage primary mucinous adenocarcinoma of the vagina. She had no history of in utero exposure to diethylstilbestrol. A digestive endoscopy and a colonoscopy were normal. She was treated by three courses of neoadjuvant chemotherapy with tri-weekly paclitaxel and carboplatin which were partially effective. However, the disease progressed rapidly and the patient died four months after the treatment.

Conclusion: Recognition of this rare entity is important and additional data about patients with primary mucinous carcinoma of the vagina should be collected and analyzed to better characterize these tumors and to elucidate its prognostic factors, optimal treatment and outcome.

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INTRODUCTION

Vaginal cancer is a rare tumor that accounts for approximately 2% of all gynecologic malignancies. Squamous cell vaginal cancers account for 80 to 85% and adenocarcinoma account for 13 to 15% of the cases (Gallup *et al.*, 1987; Platz, 1995). There are subtypes of adenocarcinoma such as papillary, mucinous adenocarcinoma and clear cell adenocarcinoma. Primary mucinous adenocarcinoma of vagina is one of the rarest subtypes. This tumor is characterized by aggressiveness and poor prognosis because of its rapid growth and recurrence, its frequent distant metastases, and its relative resistance to conventional treatment modalities including surgery, radiotherapy and chemotherapy. We report a case of advanced stage of primary mucinous adenocarcinoma of the vagina, diagnosed in our service in January 2015, showing a highly aggressive course.

Case report

A 35 year old woman presented with chief complaint of per vaginal bleeding since six months. She had no history of in utero exposure to diethylstilbestrol. On examination, abdomen was soft with no palpable mass or organomegaly. On local examination labia majora and minora were normal. Per speculum examination showed an ulcer in the lower one third of vagina on the right lateral wall extended to the anterior wall.

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The ulcer measured approximately 4 × 3 cm in size. Posterior and left lateral walls of vagina were free. Cervix and the fornices looked normal. On per vaginal examination, uterus was normal in size. No mass felt in the fornices. Bimanual per rectal examination showed that rectum was free from the lesion. No palpable lymph nodes found. Biopsies of the ulcer and of the cervix were taken. After an immunohistochemical examination, a diagnosis of mucinous adenocarcinoma of the vagina was rendered. The biopsy of the cervix was negative. An upper gastro-intestinal endoscopy and a colonoscopy were normal. On biology, there were high levels of carcinoembryonic antigen and carbohydrate antigen 125 with values of 316.9ng/ml and 100.8U/ml, respectively. The Carbohydrate antigen 19-9 was normal with a value of 2.28 U/ml. Thoracic and abdomino-pelvic CT scan showed a pelvic mass measuring 67 mm long and multiple peritoneal nodules suggestive of a peritoneal carcinomatosis, as well as a left inguinal node of 27 mm. The patient was treated by three courses of neoadjuvant chemotherapy with tri-weekly paclitaxel and carboplatin which were partially effective. However, the disease progressed rapidly and the patient died four months after the treatment.

DISCUSSION

Primary mucinous adenocarcinoma of the vagina is a rare entity. This cancer may affect a wide age range of adult women.



**Figure 1. Adenocarcinoma of the vagina:
CT scan showing a left pelvic mass**

The majority of vaginal carcinomas are metastatic tumors. The primary site of metastatic vaginal carcinoma is most frequently the uterus. Primary invasive vaginal carcinoma accounts for approximately 1.2% of all gynecological malignancies. Of these, 80 to 85% are squamous cell carcinomas, with only 13 to 15% being adenocarcinoma (Gallup *et al.*, 1987; Platz, 1995). A primary site from the cervix or endometrium must be always excluded. In our case a cervix biopsy was negative and a primary digestive site was also excluded. In a majority of cases, there are no risk factors or causes identified for Primary mucinous Adenocarcinoma of Vagina. The histogenesis of such tumors is not yet well understood. Some tumors have been reported to arise in vaginal adenosis of Diethylstilbestrol-unrelated patients (Yaghsezian *et al.*, 1992). The signs and symptoms of Primary mucinous Vaginal Adenocarcinoma include dysuria, dyspareunia, and the presence of a mass in the vagina. There are no official treatment guidelines for vaginal cancer, and most published data are derived from small retrospective studies. The treatment of Primary mucinous Adenocarcinoma of Vagina involves surgery, which is the most common treatment option considered. It can be a vaginectomy, a radical trachelectomy, or a pelvic exenteration. The location of disease, the size of the lesion, and the clinical stage of the tumor should help guide treatment planning. Stage I and II disease with squamous cell lesions at the apex or the upper posterior or lateral portions of the vagina may be treated surgically.

External-beam radiation therapy (EBRT) is recommended in patients with stage I poorly differentiated tumors and deeply invasive lesions and in all patients with stage II-IV disease. Surgical management that does not result in adequate margins mandates adjuvant radiation therapy. Concurrent cisplatin-based chemotherapy should be considered in conjunction with radiation therapy (Samant *et al.*, 2007). Prognostic factors are stage of the disease at the time of diagnosis and type of the lesion. An early diagnosis and prompt treatment of the tumor generally yields better outcomes than a late diagnosis and delayed treatment. Primary mucinous adenocarcinoma of vagina is characterized generally by aggressiveness and poor prognosis because of its rapid growth and recurrence, its frequent distant metastases, and its relative resistance to conventional treatment modalities (Nasu *et al.*, 2010). The patient in this case had an advanced stage of the disease, thus early diagnosis is very important so that effective treatment can be done with good quality of life. More aggressive treatments, possibly including novel systemic biologic agents, may be needed to achieve acceptable cure rates.

Conclusion

Because of its rarity, little is known about the etiology and the behavior of primary mucinous adenocarcinoma of the vagina. Additional data about patients with this rare tumor should be collected and analyzed to better characterize these tumors and to elucidate its prognostic factors, optimal treatment, and outcome.

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