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Case Report

CASE REPORT

Primary vaginal adenocarcinoma of intestinal-type: case report of a rare gynaecological tumour

Aloy O. Ugwu[1], Muibat Haruna[2], Kehinde S. Okunade[3,*,†], Ephraim Ohazurike[4], Rose I. Anorlu[5] and Aina A. F. Banjo[6]

[1]Department of Obstetrics and Gynaecology, Lagos University Teaching Hospital LUTH, Idi-Araba, Lagos, Nigeria, [2]Department of Anatomic and Molecular Pathology, Lagos University Teaching Hospital LUTH, Idi-Araba, Lagos, Nigeria, [3]Department of Obstetrics and Gynaecology, Lagos University Teaching Hospital LUTH, Idi-Araba, Lagos, Nigeria, [4]Department of Obstetrics and Gynaecology, Lagos University Teaching Hospital LUTH, Idi-Araba, Lagos, Nigeria, [5]Department of Obstetrics and Gynaecology, Lagos University Teaching Hospital LUTH, Idi-Araba, Lagos, Nigeria and [6]Department of Anatomic and Molecular Pathology, Lagos University Teaching Hospital LUTH, Idi-Araba, Lagos, Nigeria

*Correspondence address. Department of Obstetrics and Gynaecology, College of Medicine, University of Lagos, PMB 12003, Idi-Araba, Lagos, Nigeria. Tel: +2348034728139; Email: kehindeokunade@gmail.com

Abstract

Vaginal cancer is rare and accounts for only 1 to 2% of all gynaecological malignancies. They arise as primary squamous cell cancers or are the result of extension from the cervix or vulva. Primary mucinous vaginal adenocarcinoma of intestinal-type is an extremely rare malignancy of unknown histogenesis with a diagnostic dilemma for the clinician and histopathologist. We presented the case of a 40-year-old Para 0^{+2} woman with the complaint of a mass in the vagina and recurrent vaginal bleeding who was evaluated and worked-up for examination under anaesthesia and biopsy of the vaginal mass. The histological examination revealed the unusual intestinal-type variant of adenocarcinoma of the vagina. Recognition of this rare entity is important, particularly to avoid the pitfall of misdiagnosing metastatic disease as primary vaginal cancer.

INTRODUCTION

Vaginal cancer is rare and accounts for only 1 to 2% of all gynaecological malignancies [1]. The vagina, however, can be a common site of metastatic gynaecological cancer, either by direct extension of cervical or vulvar tumours or through lymphatic or vascular deposits. Metastatic or direct extension of non-gynaecologic tumours to the vagina can also occur from the urinary bladder, urethra, periurethral glands, rectum, and rarely the breast, lung, or other sites [2]. Secondary or metastatic cancers of the vagina are seen more frequently than primary vaginal cancers [3]. The majority of vaginal cancers are squamous-cell in origin, followed by adenocarcinoma, melanoma, and various other rare histological types [2]. Adenocarcinoma accounts for the great majority of primary vaginal malignancies in young patients and may arise in areas of vaginal adenosis,endometriosis, Wolffian duct remnants, or periurethral glands [3, 4]. In this report, we present a primary adenocarcinoma of the vagina with the extremely rare intestinal-type variant and an overview of its associated diagnostic dilemma.

†Kehinde S. Okunade, http://orcid.org/0000-0002-0957-7389
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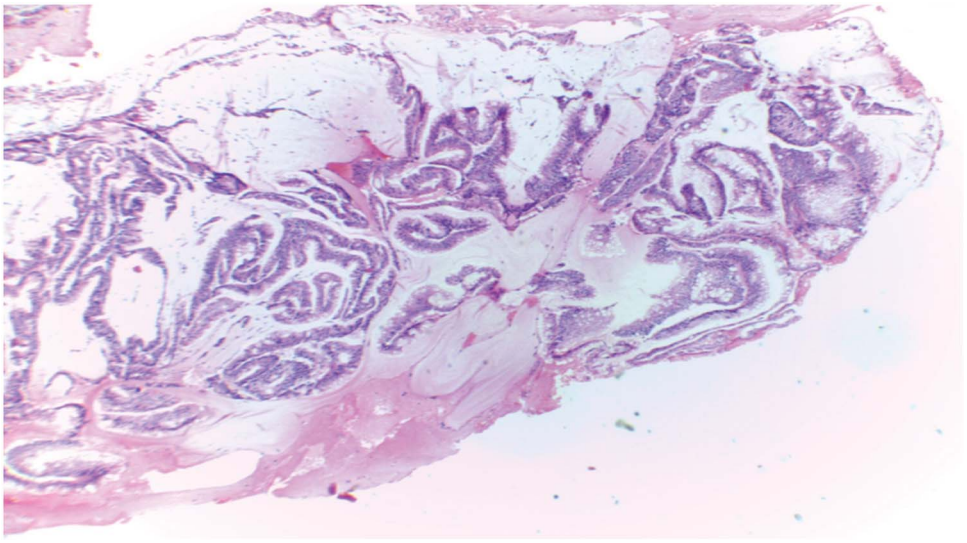

Figure 1: Photomicrograph of glands in mucin and necrotic background

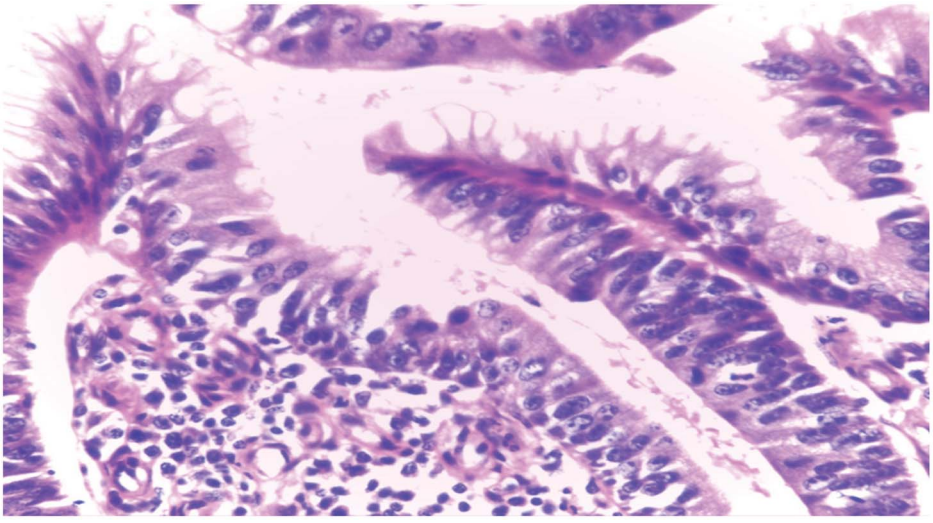

Figure 2: Photomicrograph of glands with pseudo-stratification and lining goblet cells.

CASE REPORT

A 40-year-old para 0^{+2} woman presented at the Outpatient Department of a public tertiary hospital in Lagos with a five-month history of a mass in the vagina and four-month history of bleeding per vaginam. The mass was initially the size of a pebble, but progressively increased in size until it was visible at the introitus. There was associated pain in the vagina which was described as sharp with a severity of 7 on a scale of 10. The pain was said to be aggravated by sitting or walking around and relieved by the ingestion of analgesics. There was also occasional vaginal bleeding which started a one month after the onset of the vaginal mass; bleeding was described as bright red, foul smelling, and not associated with her menstrual cycle. She had no history of chronic irritation or previous surgical instrumentation of the vagina. Physical examination findings were essentially normal; however, her vulva inspection revealed a cauliflower growth at the introitus. Digital examination showed no abdominopelvic mass and the cervix was deviated to the left but appeared normal to palpation. The adnexa was free bilaterally. Sterile speculum examination showed a cervix that was deviated towards the left lateral aspect of the vagina and appeared grossly normal. There was a friable, non-tender mass measuring about 6cmx3cm at the posterior wall of the lower-third of the vagina and extending to the introitus. The patient was counselled on the findings and the need for examination under anaesthesia (EUA) and biopsy of the vaginal mass which she consented to. Her abdominopelvic ultrasound and computerised tomography (CT) scans revealed normal sized uterus and adnexa and chest X-ray showed no abnormality. Her full blood count and serum electrolyte, urea and creatinine were all within normal limits. Retroviral screening, hepatitis B and C virus screenings were all negative. Her Pap smear was negative for intraepithelial lesion or malignancy. She subsequently had EUA and punch biopsy of the vaginal mass in theatre and the histological examination of the tissue revealed glands with widespread atypical pseudostratified cuboidal to columnar cells and goblet cells floating in a mucinous background and with the cells lining containing pleomorphic, hyperchromatic nuclei and vacuolated cytoplasm in areas with an intestinal-type appearance

[Figs 1 and 2]. A diagnosis of invasive adenocarcinoma (intestinal-type mucinous carcinoma) was made. Patient was managed as FIGO clinical stage 1 disease and she was subsequently referred to the Radiation Oncology unit for chemoradiation therapy which comprised of intracavitary/external beam radiation therapy combined with weekly dose of intravenous Cisplatin. Patient responded optimally to the treatment and she was still on follow-up as at the time of writing this report.

DISCUSSION

Primary vaginal cancer is rare. Most of these tumours are squamous cell carcinomas, others are adenocarcinomas, sarcomas and melanomas [5]. Primary vaginal cancers arise in the vagina and do not involve the cervix superiorly or the vulva inferiorly [6]. Approximately 5% of primary vaginal malignancies are adenocarcinomas. Several subtypes of these vaginal adenocarcinomas have been reported in the literature and they include; clear cell, endometrioid, serous, and mucinous carcinomas [7]. The most common variant is the clear-cell adenocarcinoma, which can occur spontaneously and in women with in-utero exposure to Diethylstilboestrol [7]. Mucinous adenocarcinomas of the vagina are extremely rare and can be further sub-classified as endocervical and intestinal types [7]. The histogenesis of vaginal adenocarcinoma of the mucinous type remains unclear. Endocervical-type mucinous adenocarcinomas have been proposed to arise from vaginal adenosis and/or endocervicosis whereas the intestinal types, which histologically resembles mucinous colonic carcinomas [8], have been reported to arise from vaginal tubular or villous adenomas and adenosis or even dysplastic enteric epithelium secondary to surgical manipulations [7, 9]. Intestinal-type adenocarcinoma of the vagina is still a challenge for the histopathologists as the tumour morphology together with the immunohistochemical stains of the tumour resembles closely that of a gastrointestinal adenocarcinoma [8]. For primary vaginal adenocarcinoma of the intestinal-type, an extensive investigation should be performed to exclude a primary adenocarcinoma of another location such as the rectum, colon, breast, ovary, uterus or cervix [8]. In this case report, examination under anaesthesia, abdominopelvic ultrasound and CT scan and chest X-ray were performed. These investigations revealed no other primary adenocarcinoma. Supplementary investigations that may also be performed if available and affordable are gastroscopy, colonoscopy, proctosigmoidoscopy, PET, MRI, cystoscopy and serum CEA level [7]. Despite its absence in our histopathogical findings, skene duct metaplasia in association with invasive adenocarcinoma had lend support to the origin of vaginal mucinous adenocarcinomas of intestinal-type to be due to metaplasia in some cases [7, 10], however, the confinement of the lesion in our patient to the posterior vaginal wall, to a large extent, excluded this. Subsequent immunohistochemical stains of the biopsied lesion, if available, is essential in this regard. The mainstay of treatment for vaginal adenocarcinomas of all variants including the intestinal type is radiation therapy with external beam radiation and/or brachytherapy, as well as surgical excision in carefully selected cases [8]. Chemotherapeutic agents are being developed both alone or in conjunction with radiotherapy for advanced cases or recurrent disease. However, due to the rarity of the disease, there have been no randomized prospective trials to guide treatment decisions [8]. There are currently no data in the literature regarding the long-term survival of these patients, however, close follow-up is recommended during the first 3 years after their primary treatment.

Intestinal-type adenocarcinoma is a rare variant of vaginal cancer with only a few reported cases in the literature. Recognition of this rare entity is important, particularly to avoid the pitfall of misdiagnosing metastatic disease as primary vaginal cancer.

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CONFLICT OF INTEREST

None declared.

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ETHICAL APPROVAL

Ethics approval for this report was obtained from the human research and ethics committee of the public tertiary hospital where the patient was being managed (ADM/DCST/HREC/APP/2644).

CONSENT

The authors certify that they have obtained all appropriate patient consent. In the consent form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understood that all her identifiable information will not be published.

GUARANTOR

The corresponding author (KSO) will be the guarantor for this paper.

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