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Clear Cell Adenocarcinoma of the Young Woman's Cervix: About A Case

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Abstract **Case Report**

This report presents a case study of a patient diagnosed with clear cell adenocarcinoma of the cervix without a history of in utero exposure to diethylstilbestrol (DES). It is important to note that despite being an uncommon occurrence, clear cell adenocarcinoma should be considered a potential diagnosis for young women with cervicovaginal lesions, even if they have no prior history of in utero DES exposure. We present a case report of a 33-year-old woman who sought medical attention for menometrorrhagia, leucorrhoea, and the discovery of a vegetative tumour. The patient was diagnosed with clear cell carcinoma of the cervix. The patient was treated with concurrent radiochemotherapy combined with endocavitary uterovaginal brachytherapy and showed a favorable progression during a six-month follow-up period.

Keywords: Clear cell adenocarcinoma, cervix, young woman, chemoradiotherapy.

I. INTRODUCTION

Clear cell adenocarcinoma (CCAC) of the cervix is a rare disease that accounts for only 4% of all adenocarcinomas of the cervix [6]. Its etiology and pathogenesis are not yet clear. Several studies have linked the occurrence of cervical or vaginal CCAC in young women to intrauterine exposure to diethylstilbestrol (DES), a synthetic nonsteroidal estrogenic hormone prescribed to pregnant women in several countries as a preventive therapy against abortion [4, 5]. On the other hand, many cases of cervical or vaginal CCAC have been reported in young women with no history of intrauterine exposure to DES [1].

CCAC often manifests as irregular vaginal bleeding. In young adult women who have irregular bleeding, the possibility of a vaginal or cervical tumor should be considered in the differential diagnosis [7]. The most important prognostic factors for CCAC are stage, tumour size and lymphovascular invasion. The most common extra-pelvic sites of relapse are the lung, liver and bones. CCAC also has a lower 5-year survival rate than carcinomas epidermoids [8]. Due to the low incidence of CCAC, there are limited data on clinical behaviour, disease characteristics, optimal management, patterns of metastasis and recurrence. We report the case of a young woman who has clear cell adenocarcinoma of the cervix followed at the oncology-radiotherapy department of the Mohammed VI University Hospital in Marrakech.

II. CASE REPORT

This is a young patient of 33 years, married mother of 4 children, without particular history who presented to her gynecologist for leucorrhoea and meno-metrorrhagia. The gynecological examination finds a 7 cm vegetative tumor infiltrating the vagina. The pathological study of the cervical biopsy revealed invasive clear cell adenocarcinoma. A pelvic MRI was performed objectifying a lesional process centered on the cervix extended to the isthmian region (Fig 1a, b) measuring 5.7x7.3x5.8 cm infiltrating two-thirds of the vagina at the bottom pushing back the rectum with respect for the fatty border of separation and integrity of the rectovaginal septum, in front it pushes back the bladder with respect for the separation border; discreetly infiltrating the parameters bilaterally that can be classified FIGO IIB. An extension assessment made of a thoraco-abdomino-pelvic CT scan was performed, revealing no secondary location.

The patient was treated with concurrent radiochemotherapy of 45 Gy in 25 fractions of 1.8 Gy in combination with endocavitary uterovaginal brachytherapy of 7 Gy x 4. At six months follow-up, the disease had not progressed.

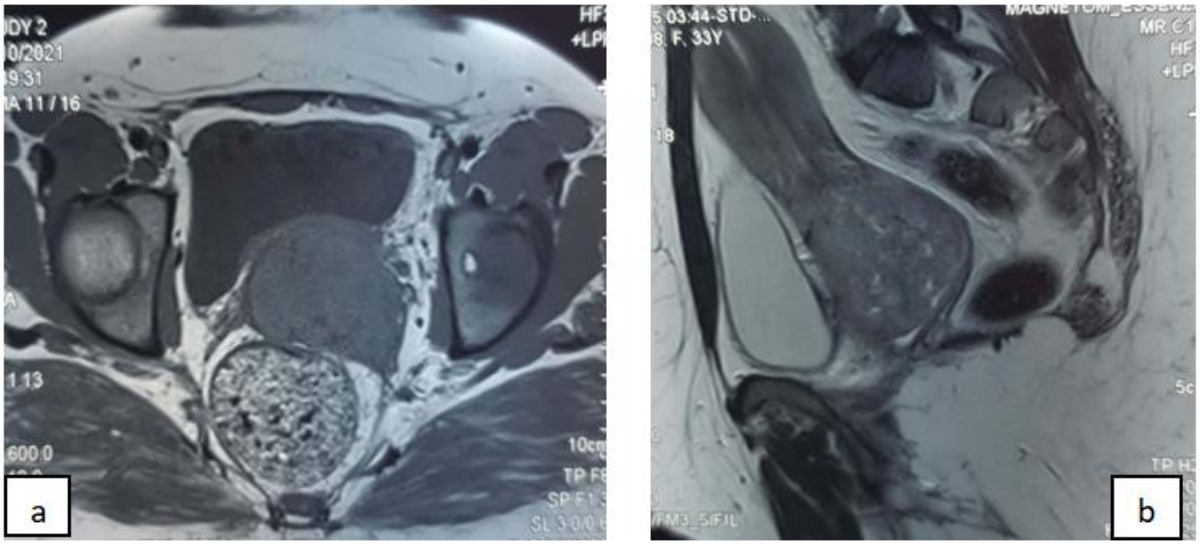

Fig 1a, b: Magnetic resonance imaging (MRI) showed a lesional process centered on the cervix extended to the isthmian region measuring 5.7x7.3x5.8 cm infiltrating two-thirds of the vagina at the bottom pushing back the rectum with respect for the fatty border of separation and integrity of the rectovaginal septum, in front it pushes the bladder with respect for the border of separation. a Transverse. b Sagittal

III. DISCUSSION

In recent years, there has been an increase in the frequency of cervical adenocarcinomas, mainly due to a decrease in invasive squamous cell carcinomas in countries where cervical neoplasia screening has been effectively implemented.

CCAC is a rare tumour, which usually occurs in young women. Hanselaar et al reported that the CCAC has a bimodal age distribution. The first peak in average age was 26 years (17 to 37), and the second was 71 years (44 to 88) [1]. Contrary to these results, in a study conducted by Seki *et al*., [9] involving 32 Japanese patients, CCAC was observed in all age groups (mean age 50.8 years). The youngest case of clear cell vaginal carcinoma reported in the literature was a one-year-old child [8].

In utero exposure to DES was a known predisposing factor of the CCAC. Recently, several cases not associated with DES have been reported after they were banned. Pirog *et al*., [2] reported the incidence of HPV infection in 760 cases of cervical adenocarcinoma, and the clear cell type had a lower prevalence of HPV at 20%. New cases of CCAC of the cervix and vagina continue to occur, often without a history of exposure to DES, as in this case [11]. In our case, the patient had no notion of in utero exposure to DES and the HPV test was not done due to lack of resources.

Treatment for CCAC is not well defined and relies heavily on the methods used to treat squamous cell carcinoma and unclear cell adenocarcinoma. In the early stages, surgery is an option. There is growing interest in fertility-preserving treatments. In more advanced stages, surgery is not recommended, as it is unlikely to be curative. In addition, advanced stages require adjuvant chemotherapy and radiotherapy, and if patients have undergone surgery, it is associated with a higher risk of complications. In our patient, we did not perform surgery because of her advanced stage radiochemotherapy was decided chemotherapy of choice is usually cisplatin combined with radiotherapy. One study also suggested that there was increased activation of the EGFR-PI3K-AKT-mTOR pathway in CCAC and that tyrosine kinase and AKT-mTOR inhibitors could be novel therapeutic targets [3].

IV. CONCLUSION

CCAC remains a rare disease without exposure to DES. In the absence of clear guidelines for the management of CCAC, more randomised controlled trials are needed to find the appropriate treatment regimen. But the rarity of the condition could prove to be a challenge in conducting large-scale trials.

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