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Didelphys Uterus with Cervical Cancer: What about Herlyn-Werner-Wunderlich Syndrome? A Case Report and Review of Literature!

ELBARAKA Yassine[1],
[1]Gynecological obstetrics department,
hospital university Hassan II,
FEZ.MOROCCO

ACHKIF Salaheddine[1]; SAOUD karam, MAMOUNI Nisrine[1], ERRARHAY Sanaa[1], BOUCHIKHI Chahrazed[1], BANANI Abdelaziz[1]
HAJJAR Chaymae[2], Maâroufi Mustapha[2].
[1]Gynecological obstetrics department, hospital university Hassan II, FEZ.MOROCCO
[2]Radiology department, hospital university Hassan II, FEZ

Abstract:- Müllerian tract anomalies are defined as deviations from normal anatomy resulting from embryological mal development of the Müllerian system or paramesonephric ducts. The Herlyn-Werner-Wunderlich syndrome (HWWS) is one of these anomalies, it associated didelphys uterus, unilateral obstructed hemivagina, and ipsilateral renal agenesis. Association between cervical cancer and HWWS is an uncommon condition in clinical practice. We present a case of a 65-year-old female, gravida 11, para 7,with no symptoms in young adulthood or teenager in relation to didelphys uterus, was admitted with atypical genital bleeding that had continued for seven months. Surgical treatment, with systematic pelvic and para aortic nodal dissection, with poor prognostic factors, she is chemoradiotherapy treatment. The primary surgical treatment is encouraged, because we can get prognostic factors and search for other congenital malformation.

Keywords:- *Müllerian duct anomalies; uterine didelphys; ipsilateral renal agenesis; obstructed hemivagina; epidermoid carcinoma, cervical cancer.*

I. INTRODUCTION

Cervical cancer is the fourth most common cancer in women[1]. Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare form of uterovaginal duplication with three characteristic anomalies, namely, didelphys uterus, unilateral obstructed hemivagina, and ipsilateral renal agenesis [2].In this case report, we discuss a rare case of a woman with Herlyn-Werner-Wunderlich syndrome who presented an epidermoid carcinoma of cervix of uterus.

II. CASE REPORT

A 65-year-old female, gravida 11, para 7, was admitted with atypical genital bleeding that had continued for seven months. On pelvic examination two vaginal cavities were noted. In the speculum, on the left, there was a tumor cervix with bleeding from the endocervix and vaginal touch founded an indurated and irregular cervix. On the right, the cervix was normal. Cytologic and histologic examination of the cervix revealed well-differentiated non-keratinizing squamous cell carcinoma. An MRI was performed, showing a bi-cervical bicornal uterine.

Malformation that each one has its own vaginal cavity. The right uterine horn presents with a 27 mm cervical tumor of great vertical and transverse axis which breaks the cervical stroma forward and which does not show an isthmic extension at the top; vaginal bottom and laterally parametrial (figure 1).Her FIGO clinical stage was Ib. A radical enlarged colpohysterectomy was performed (figure 2).As pathologic examination of the resected specimen of right uterusdisclosed Well differentiated epidermoid carcinoma mature and infiltrating the entire cervix (figure 3) circumferentially and extending to the isthmus without infiltration of parameters.

III. DISCUSSION

Müllerian duct anomalies are congenital defects of the female genital system that result from agenesis of defective vertical or lateral fusion, canalization, or resorption failure of the Müllerian or paramesonephric ducts. It occurs between 6 and 22 weeks in utero [3].The Herlyn-Werner-Wunderlich syndrome was initially described by Herlyn and Werner then by Wunderlich. It is also knownas OHVIRA syndrome (obstructed hemivagina and ipsilateral renal anomaly).It is a rare Müllerian duct anomalycharacterized by uterus didelphys, unilateral obstructed hemivagina and ipsilateral renal anomalies. Itsprevalence is between 0.1%–3.8% [4].Furthermore, just few casesof carcinoma of the lower genital tract at patient with genital tract malformations, were reported. It is due probably to the difficulty of diagnosis[5]. Beside this, according to Zonget al., patients with genitourinary tract malformations had a high probability of presenting with adenocarcinoma of the lower genital tract [6]. There were 2 cases of adenocarcinoma and one case of carcinosarcoma associated to HWWS [8]. There were also few cases of epidermoid carcinoma associated to didelphys uterus [9-10]. But, to our knowledge, there is no case that has reported an epidermoid carcinoma associated toHWWS,so our case is the first

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described. The principal and common symptom of cervical cancer andof genital tract malformations in some patients, but this one can be totally asymptomatic [12,13], what was the case of our patient, but what can explain more this delayed diagnosis although our patient wasgravida 11, para 7, is the absence of monitoring of her pregnancies. In the other side, it seems that the diagnosis of carcinoma of the lower genital tract is more difficult in women with genital tract malformations than in those without such malformations [6]. The imaging of choice in Müllerian anomalies isan MRI, its accuracy and better resolution on soft tissue characterize better uterus and vagina.It is also the imaging of choice to evaluate and staging cervical carcinoma. However, ultrasonography is previously done,or 3D ultrasound whichmake the diagnosis [14,15]. The aim of surgical and medical management for HWWS is not just to provide symptomatic relief, but also to prevent complications, and to maximize the fertility of a patient. The management of epidermoid carcinoma in a patient with HWWS needs a multidisciplinary team composed of an obstetrician-gynecologist, radiologist, psychologist, nephrologist, and oncologist, and the decision making needs to be precise and personalized. In patientswith Müllerian duct anomalies and cervical cancer, clinical staging can be ambiguous, and the common association with renal agenesis in HWWS could influence the use of potentially nephrotoxic agents, like cisplatin [11]. In literature, the lymphatics and nodes of the various Müllerian ducts anomalies are not described, That is why, in a patient who has Müllerian anomalies with cervical cancer, radical surgery, a lymphatic node dissection pelvic and retroperitoneal in stage IIB or advanced is recommended. The surgical approach provides the real stage and improves radiotherapy field treatment with more success and less morbidity [8-9]. Finally, there were no studies comparing the surveyof cervical cancer in patients with Müllerian tract anomalies to other patients without those anomalies.

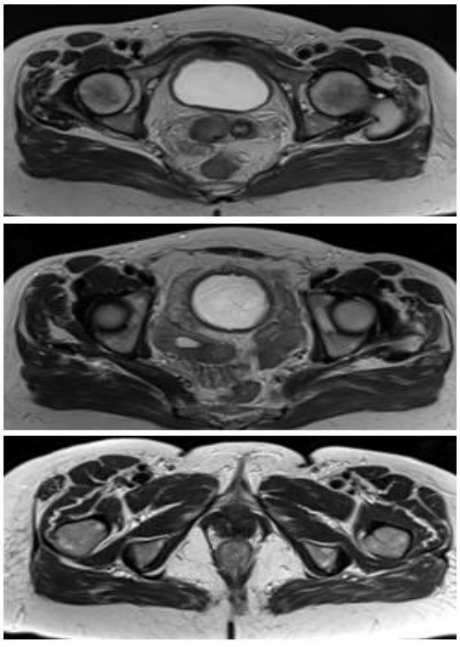

Figure 1: MRI in axial sections, objectivizing 02 vaginas, 02 cervixs and 02 uterine cavities.

IV. CONCLUSION

Cervical cancer, including epidermoid carcinoma, can also touch patients with Müllerian tract anomalies, like Herlyn-Werner-Wunderlich syndrome. In fact, it is preferable that will be more studies which can inform and help practitioners to the management of this association.

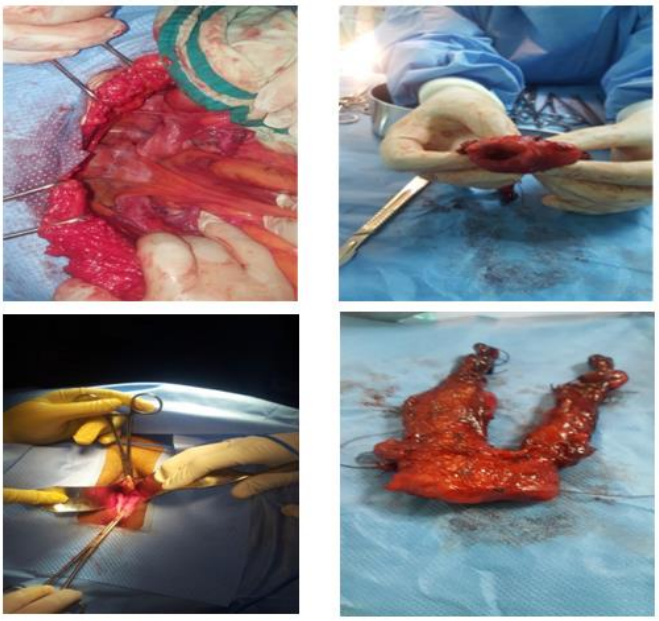

Figure 2: Intra-operative photos showing didelphys uterus with the tumor on the left cervix

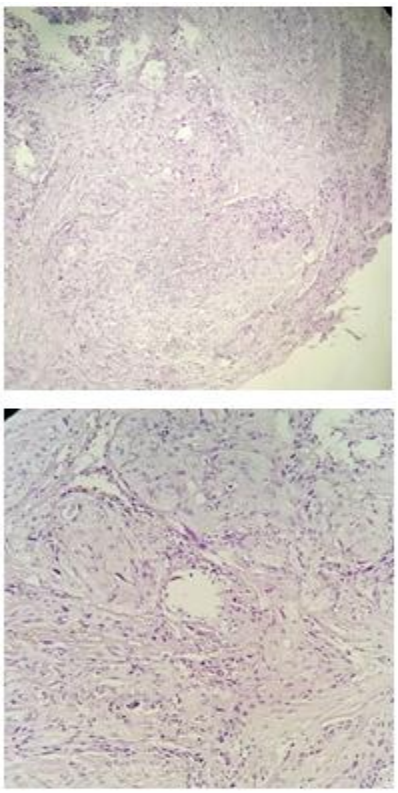

Figure 3: large tumour cells with atypical nuclei and surrounded by an abundant eosinophilic cytoplasm with clear cytoplasmic limits, arranged in massifs.

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