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Case Report

Primary vulvovaginal choriocarcinoma: a case report of unusual presentation and literature review

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ABSTRACT

Only one case of primary extra uterine vaginal choriocarcinoma and one case of primary vulvar choriocarcinoma have been reported in literature. This is a case of 27 year old lady who presented with a 10cm × 7cm × 5cm vulvar mass with pain abdomen since 1 month, to the Gynecologic oncology outpatient. The mass was smooth, hard and fixed to underlying structures. Multiple bilateral inguinal lymph nodes were enlarged. Vulvar biopsy with Immunohistochemistry proved it to be choriocarcinoma. CT scan thorax, abdomen and pelvis showed multiple bilateral lung metastases, empty uterine cavity and normal sized uterus with a vaginal mass extending up to introitus encasing urethra and anal canal with multiple enlarged pelvic & inguinal lymph nodes. Final diagnosis of Primary Vulvovaginal choriocarcinoma FIGO stage III and WHO score-12 was made. Multidrug chemotherapy with Etoposide, Methotrexate, Actinomycin-D, Folinic Acid, Cyclophosphamide and Vincristine (EMA-CO) was started then shifted to Etoposide, Methotrexate, Actinomycin-D, Folinic Acid and Cisplatin (EMA-EP) regimen followed by Paclitaxel & Carboplatin, because of poor response. Patient's βHCG became 1.57IU/L with resolution of all lesions after 5 three weekly cycles of Paclitaxel & Carboplatin. Now she is planned for three more cycles of chemotherapy. This case highlights another atypical presentation of choriocarcinoma.

Keywords: Vulvar, Vaginal, Choriocarcinoma

INTRODUCTION

Primary extra uterine choriocarcinoma is very rare. The diagnostic criteria for this entity are as follows a) absence of disease in the uterine cavity b) pathologic confirmation of the diagnosis c) exclusion of molar pregnancy d) exclusion of a coexistent normal intrauterine pregnancy.³ Though there are several cases of uterine choriocarcinoma metastatic to vulva or vagina⁴⁻¹⁰, only one case of primary extra uterine vaginal choriocarcinoma and one case of primary vulvar choriocarcinoma have been reported in literature.^{1,2} Here we present a case of 27 year old woman who presented with a hard vulvar mass 5 months after abortion.

CASE REPORT

A 27 year old woman was admitted in the Department of Gynecologic oncology with constant dull aching pain in lower abdomen since one month with a 10×7×5cm vulvar mass on left side (Figure 1).

The vulvar mass was slow growing and painful. She was having difficulty in micturating and defecating with sense of incomplete evacuation. Her last delivery was 3 years back but 5 months before she had aborted a 3 month pregnancy spontaneously & completely without any medical aid. This was followed 4-6 weeks later by regular 28-30 days cycle lasting for 4-5 days with average flow, her last period was approximately 10 days before she presented to the hospital. On examination she had a body

mass index of 16.8kg/m². Bilateral multiple inguinal lymph nodes were palpable, mobile, 1-2 cm in size. Liver was enlarged 2-3cm below right costal margin. The vulvar mass was hard, fixed, tender, involving the left labium majus and to some extent right labium majus, with smooth surface. Vaginal or rectal examination could not be done because of the hard, tender & fixed mass encroaching. A provisional diagnosis of soft tissue sarcoma or Bartholine gland tumor was made.



Figure 1: Initial vulvar mass.

Chest X ray showed multiple well defined soft tissue opacity in both lung fields suggestive of lung metastases. Vulvar biopsy was taken after blood transfusion as hemoglobin was 6gm/dl at the time of admission. There was slight bleeding at the biopsy site which stopped after local application of pressure for few minutes. Vulvar biopsy histopathology report was suggestive of undifferentiated tumor with possibilities of germ cell tumor, undifferentiated cancer or melanoma. The Immunohistochemistry report was HCG, AE-1, CK-7, CD-10 positive and viamentin, EMA, PLAP and S-100 negative with the final diagnosis of choriocarcinoma (Figure 2). The USG pelvis showed endometrial thickness of 6mm; uterus was normal size & echo pattern and a space occupying lesion in vagina. MRI brain was normal. CT scan thorax showed multiple bilateral lung metastases largest lesion measuring 61×50mm on right side and 34×30mm on left side. CT abdomen, pelvis (Figure 3) showed "Uterus without any abnormal enhancing lesion within, multiple heterogeneously enhancing enlarged necrotic nodes along both external & internal iliac vessels, enlarged bilateral obturator & inguinal nodes, enlarged iliac nodes on right side were noted infiltrating the iliacus muscle. Multiple conglomerated peripherally enhancing soft tissue lesions were noted along bilateral vaginal walls. The lesion reached up to anal verge encasing it with loss of fat plane. It encased the urethra. Hepatomegaly with slightly enlarged spleen was present without any space occupying lesion. Her βHCG value was 2, 48,844IU/L, CEA - 18.62ng/ml and Serum LDH was 1688Unit/liter. She was thus diagnosed to have Primary Vulvovaginal choriocarcinoma, FIGO stage III with modified WHO score of 12. She was started on Etoposide, Methotrexate,

Actinomycin-D, Folinic Acid, Cyclophosphamide and Vincristine the so called EMA-CO regimen and given till 3 cycles when the βHCG values started to rise on weekly estimation from 269.5IU/L, 360.2IU/L to 487IU/L. She was then shifted to EMA-EP regimen (Etoposide, Methotrexate, Actinomycin-D, Folinic Acid and Cisplatin) which was given for 2 cycles after which βHCG values rose from 146.1IU/L, 209.3IU/L to 852.9IU/L on weekly estimation so patient was shifted to 3 weekly Paclitaxel (175mg/m²) & Carboplatin (AUC 5) regimen. Patient responded to this, the vulvar and vaginal lesion disappeared βhCG returned to 1.57IU/L (Normal <5IU/L) after 5 cycles of Paclitaxel & Carboplatin and other tumor markers became nondetectable. Now 3 more cycles of the same chemotherapy is planned for the patient.

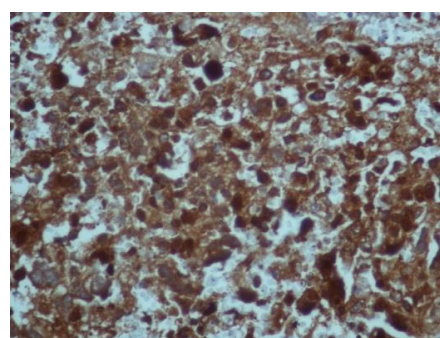


Figure 2: 200X view of βHCG positive vulvar biopsy specimen.



Figure 3: CT scan pelvis showing vaginal lesion (Red arrow).



Figure 4: Reduced size of lesion after 2 cycles of Paclitaxel + Carboplatin.

DISCUSSION

Primary extra uterine genital choriocarcinoma may be gestational as a complication of pregnancy or nongestational of germ cell origin. The difference between the two can be made definitely only by DNA polymorphism.¹¹ The circumstantial evidence of reproductive age with recent history of abortion and prompt response to chemotherapy in this case however point towards gestational origin of the tumor.² There are some hypothesis regarding uterine noninvolvement in cases like this, like resolution of uterine tumor after metastases to other sites or probability of missing the uterine pathology on USG and CT scan.^{1,2} Absence of vaginal bleeding or menstrual complaints in this case is explained by uterine noninvolvement.

PubMed, Proquest, Google and Embase databases were searched for similar cases with the terms like choriocarcinoma sites, extra uterine/vulvar/vaginal/ectopic/gestational/nongestational choriocarcinoma etc. and only one case of primary vaginal choriocarcinoma was found reported in 1976 in a postmenopausal lady.¹ She presented with vaginal bleeding and coughing. One case of primary vulvar choriocarcinoma was reported by Weiss S. in 2001.² Their case was a 31 year lady presenting with vaginal bleeding and vulvar lesion developed 1-2 months after initial presentation.

CONCLUSION

Though it is not realistic to keep choriocarcinoma as differential diagnosis in such vulvar lesion, it should be kept in mind before taking biopsy when the lesion has a recent and rapid history of development & is accompanied by multiple lung metastases. It was fortunate in this case that there was only minimal bleeding after biopsy otherwise choriocarcinoma is notorious for uncontrolled hemorrhage.

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