Magnetic Resonance Imaging of a Rare Case of Primary Choriocarcinoma of the Vulva

Sathyanathan, Babu Peter¹ and Mridhula, Venkatakrishnamuruali Maheswari¹

¹Barnard Institute of Radiology, Madras Medical College, Chennai, Tamil Nadu, India

Abstract

Gestational choriocarcinoma usually occurs in the corpus uteri in association with a coincident or antecedent pregnancy (1). Primary extrauterine choriocarcinoma is a rare entity, with the cervix being the most common site (2). We report a rare case of primary choriocarcinoma of the vulva in a woman with previous partial molar pregnancy and highlight its Magnetic Resonance Imaging (MRI) features.

Keywords: Gestational Choriocarcinoma, Vulva, Magnetic Resonance Imaging (MRI), Gestational Trophoblastic Neoplasia (GTN)

Patient and Observation

The patient was a 26 years old female who presented with complaints of menorrhagia. She had undergone suction evacuation for partial molar pregnancy 4 months ago, which was confirmed by histopathological examination. Following an initial decrease in serum Beta human chorionic gonadotropin (β-hCG) in the first month following the procedure, she had failed to follow up and the β-hCG level at the time of presentation was 30,292 IU/L. Magnetic Resonance Imaging (MRI) of the pelvis was done using 1.5T MRI (AVANTO Siemens Healthineers, Erlangen, Germany). Standard institute protocol was followed, signal acquisition was done using phased array body coil. T1 sagittal, T2 axial, sagittal and coronal, 3D SPACE T2, Short Tau Inversion Recovery (STIR) coronal, T1 weighted fat suppressed axial and dynamic contrast enhanced images and axial diffusion weighted imaging (DWI) and gradient (GRE) sequences were used. These revealed a well-defined mass lesion in the right sublabial region, measuring 3.6 x 2.8 x 2.4cm in its greatest dimensions. On T1 weighted images, the lesion was predominantly hypointense, with few internal hyperintensities. On gradient images, these T1 hyperintensities showed blooming, indicating that these were areas of hemorrhage. On T2 weighted images, the lesion was heterogeneous, with predominant hyperintensity and a T2 hypointense rim. Few flow voids were noted in the center of the lesion. Few peripheral areas of restricted diffusion were noted on DWI. The uterus and adnexa appeared normal. On contrast administration, there was intense peripheral enhancement of the lesion, especially in the regions of restricted diffusion. Intense enhancement was also noted in the center of the lesion, where flow voids were observed on T2 weighted images. Few confluent non enhancing areas which were hyperintense on T2 weighted images were also observed, indicative of necrosis. There was no evidence of invasion of the vaginal wall or enlarged pelvic lymph nodes (Fig. 1-4).
Chest X ray was normal. In view of the clinical presentation, elevated β-hCG and imaging findings, a diagnosis of choriocarcinoma of the vulva was made and she was started on chemotherapy (EMA-CO regimen – Etoposide, Methotrexate, Actinomycin, Cyclophosphamide, Vin-cristine). β-hCG decreased progressively to 464 IU/L following the first cycle and subsequently, to 0.32 IU/L following the fifth cycle. The patient is currently asymptomatic. Follow up MRI was done to check for any residual lesion or sequelae. This revealed a normal uterus (Fig.5) and adnexa with no abnormal significant residual lesion in the vulva.

**Discussion:**

Gestational choriocarcinoma is a malignant form of Gestational Trophoblastic Neoplasia (GTN) and can be associated with any type of coincident or antecedent pregnancy (1). Extrauterine choriocarcinoma is extremely rare and the cervix is the commonest site (2). Only four cases of primary vulvovaginal choriocarcinoma and two cases of primary vaginal choriocarcinoma are found in literature, with imaging features described only for purely vaginal lesions (3,4,5,6,7,8). The commonest presenting feature is abnormal vaginal bleeding with elevated β-hCG, as seen in our case (2). These lesions are usually hypointense on T1 weighted images with internal areas of hyperintensity indicating hemorrhage. On T2 weighted images, they are heterointense with a hypointense rim. On contrast administration, they exhibit peripheral enhancement with non-enhancing areas indicating necrosis (7,8). The imaging features in our case are similar to all the features thus described. These masses are highly...
Figure 4: Contrast enhanced T1 fat saturated sagittal (a) and axial (b) images and subtracted image (c) demonstrate peripheral enhancement (blue arrow) of the lesion with intense enhancement in the center, at the region of flow voids (yellow arrows).
vascular, as evidenced by the flow voids and avid enhancement in the center of the lesion in our case (2,7,9). Choriocarcinomatous lesions contain abundant venous plexus without valves, which makes them prone to torrential hemorrhage, due to which biopsy is not routinely advised when β-hCG is rising or elevated (10). These tumors are also highly chemo sensitive and carry a good prognosis if treated (10). In our case too, there was a significant fall in β-hCG following the first cycle of chemotherapy itself and complete resolution of the lesions post chemotherapy.

Conclusion:
Primary choriocarcinoma of the vulva has rarely been reported and review of literature revealed no case with MRI features has been reported till date for choriocarcinoma involving only the vulva. An awareness of a lesion in the labial region being due to an extraterine choriocarcinoma in a patient with elevated β-hCG is crucial to guide management and aid in complete recovery from this potentially fatal malignancy. Our case is also unique, as we highlight the pretreatment MRI features as well as post treatment imaging follow up.

Correspondence to
Dr. Mridhula, V M
https://orcid.org/0000-0001-8903-4458
Barnard Institute of Radiology
Madras Medical College
Rajiv Gandhi General Hospital
Opposite to Chennai Central Railway Station
Chennai, Tamil Nadu, India

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