Primary gestational choriocarcinoma of the vagina: magnetic resonance imaging findings

Case report

A 33-year-old nulligravid Filipino woman was admitted to the gynaecology ward of Princess Margaret Hospital in April 2014, with lower abdominal pain and recurrent retention of urine. Her pregnancy test was positive. Per-vaginal examination revealed a 4-cm firm, fixed, and wide-based vaginal mass with smooth wall over the upper anterior vagina. Bedside transabdominal ultrasonography confirmed a 4.5-cm solid mass in the anterior vagina and a 3.5-cm intramural fibroid over the left side of the uterus. No adnexal mass or free fluid was seen. Beta human chorionic gonadotropin (β-hCG) level was elevated to 270 743 IU/L (reference level, <5.0 IU/L for non-pregnant women), and repeated checking 3 days later was 253 957 IU/L. Alpha-fetoprotein, carcinoembryonic antigen, and Ca-125 were negative. A urologist was consulted and the patient underwent flexible cystoscopy that revealed indentation of the urinary bladder by an external mass. Mild erythematous changes were visualised at the right urinary bladder base. Biopsy revealed inflammation but no malignant cells.

Magnetic resonance imaging (MRI) of the pelvis revealed a 4.8 cm x 5.2 cm x 4 cm (craniocaudal x anteroposterior x transverse) mass located at the anterior aspect of the vagina and a 3.5-cm intramural fibroid over the left side of the uterus. No adnexal mass or free fluid was seen. Beta human chorionic gonadotropin (β-hCG) level was elevated to 270 743 IU/L (reference level, <5.0 IU/L for non-pregnant women), and repeated checking 3 days later was 253 957 IU/L. Alpha-fetoprotein, carcinoembryonic antigen, and Ca-125 were negative. A urologist was consulted and the patient underwent flexible cystoscopy that revealed indentation of the urinary bladder by an external mass. Mild erythematous changes were visualised at the right urinary bladder base. Biopsy revealed inflammation but no malignant cells.

Magnetic resonance imaging (MRI) of the pelvis revealed a 4.8 cm x 5.2 cm x 4 cm (craniocaudal x anteroposterior x transverse) mass located at the anterior aspect of the vagina. It was hypointense with a faint hyperintense rim on T1-weighted images (Fig 1a) and heterogeneously hyperintense
with hypointense rim on T2-weighted images (Fig 1b). Peripheral enhancement was observed while the central part remained non-enhanced (Figs 1c and 1d). Part of the mass closely abutted the cervix and urinary bladder, where intervening fat planes could not be well-delineated. The urinary bladder wall was trabeculated. Dilated enhancing tortuous tubular structures were seen at the left adnexal region and numerous signal void foci were observed at the uterine wall. These were due to the presence of high-flow vasculatures related to tumour hypervascularity (Figs 1e to 1h). There was also an incidental finding of a predominantly T2 hypointense non-enhancing intramural fibroid at the posterior uterine fundus (Figs 1b and 1d). No ovarian mass was detected and no intra-uterine gestational sac or ectopic pregnancy.

Later examination under anaesthesia revealed that the lower part of the vaginal lesion had ruptured. Excision of part of the vaginal lesion was performed. Tissue frozen section confirmed it to be choriocarcinoma with no other germ cell component. DNA polymorphism analysis was performed in a microsatellite analysis using six markers by extracting DNA from the microdissected tumour cells, then comparing it with normal DNA extracted from a peripheral blood sample. The result favoured gestational choriocarcinoma. In addition, chest X-ray revealed multiple ill-defined opacities in both lungs, measuring about 0.8 to 1.2 cm in size (Fig 2a).

The patient was diagnosed with primary gestational choriocarcinoma of the vagina with lung metastases, and suspicious infiltration of the urinary bladder and cervix. She was further managed at the gynaecomatology specialist centre and given chemotherapy (etoposide, methotrexate, actinomycin, and cisplatinum). The β-hCG level dropped to 5604 IU/L with interval regression of the pulmonary nodules after initiation of chemotherapy (Fig 2b).

Discussion

Gestational trophoblastic disease comprises a wide spectrum of benign/premalignant to malignant conditions and includes hydatidiform mole (complete or partial), invasive mole, choriocarcinoma, and placental-site trophoblastic tumour. The incidence varies in different countries with the highest rate noted in South-East Asia. The cause of such variation is not well understood. Choriocarcinoma is a malignant form of gestational trophoblastic neoplasia, and generally arises in the uterine corpus of women of reproductive age with coincident or antecedent pregnancy. Primary extrauterine choriocarcinoma is rare and most reported cases have occurred in the uterine cervix. It is thought to arise along the path of migration of germ cells to gonads, or de-differentiated from another histological type. Our case was very unusual as it occurred in the vagina. The most common site of metastasis for choriocarcinoma is lung, and this also occurred in our patient.

The clinical diagnosis of extraterine choriocarcinoma is extremely difficult as symptoms are often non-specific. Vaginal bleeding is the most common presenting symptom, but as such often mimics other more common disease entities.
Primary extrauterine choriocarcinoma has been misdiagnosed as ectopic pregnancy, dysfunctional uterine haemorrhage, and cervical polyp. This can often lead to a delay in proper management. Since trophoblastic neoplasm such as choriocarcinoma produces an excessive amount of β-hCG, serial monitoring of its trend is very helpful for diagnosis and follow-up of treatment response. The mainstay of treatment is chemotherapy, as choriocarcinoma is highly chemosensitive. For a nulligravid woman as in this case, differentiation between gestational or non-gestational origin merely from a clinical history can be confusing. Differentiation by DNA polymorphism analysis is useful to guide the management as non-gestational choriocarcinoma is known to have a worse prognosis and to be resistant to single-agent chemotherapy.

To the best of our knowledge, the imaging findings of primary extrauterine choriocarcinoma are rarely described in the literature, particularly that of vaginal origin. In the present case, MRI showed a hypointense vaginal tumour with hyperintense rim on T1-weighted images and heterogeneously hyperintense with hypointense rim on T2-weighted images. Rim enhancement was observed on T1-weight images after gadolinium contrast injection. The non-enhancing centre of the tumour could be due to tumour necrosis, commonly seen in choriocarcinoma. Poor delineation of fat planes with cervix and urinary bladder raised the suspicion of tumour involvement. Features of chronic bladder outlet obstruction were present as suggested by the trabeculated bladder outline. The signal void foci over the uterine wall and the high-flow vasculatures at the left adnexal region signified angiogenesis and neovascularisation, thus the hypervascular nature of the tumour. This is in accordance with the characteristic hypervascularity of choriocarcinoma. The multiple nodular opacities on chest X-ray are most likely represented lung metastases, although histological confirmation was not performed. Interval shrinkage of these nodules was observed on chest X-ray following initiation of treatment.

In summary, imaging findings of a hypervascular tumour and exceedingly high levels of β-hCG are useful in making the diagnosis of extrauterine choriocarcinoma, and MRI is valuable in assessing extrauterine extension, tumour vascularity, and overall staging of the tumour.

References