Extremely Rare Case of Bilateral Pure Primary Non-Gestational Ovarian Choriocarcinoma

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Summary

Background:
Germ cell tumors of the ovary constitute less than one percent of ovarian tumors worldwide. Choriocarcinoma arising \textit{de novo} from the ovary is very rare and only occasionally reported in the literature. Herein, we report a case of bilateral non-gestational pure primary ovarian choriocarcinoma that was confirmed by beta human chorionic gonadotropin (\(\beta\)-HCG) levels and histopathology.

Case Report:
Our case is of a middle-aged multiparous female who presented with amenorrhea for three months. She underwent an evaluation with ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI), which revealed bilateral bulky solid adnexal masses. Based on an increased blood level of the beta human chorionic gonadotropin and a histopathological examination, the diagnosis of bilateral non-gestational pure primary ovarian choriocarcinoma was made.

Conclusions:
The imaging findings were found to be specific for bilateral non-gestational pure primary ovarian choriocarcinoma.

MeSH Keywords: Choriocarcinoma • Chorionic Gonadotropin • Neoplasms, Germ Cell and Embryonal • Ovarian Neoplasms

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on the gradient echo (GRE) sequence due to hemorrhage (Figure 9). Avid peripheral enhancement of both lesions was seen on post-gadolinium enhanced images (Figure 10). The uterus was separately visualized and appeared normal. Fat planes with adjacent structures were preserved. The chest radiograph, that was performed to evaluate pulmonary metastases, was unremarkable.

Subsequently, the patient underwent abdominal hysterectomy with bilateral salpingo-oophorectomy. On gross examination, both ovaries appeared bulky with few hemorrhagic cystic spaces (Figure 11). Microscopic examination was confirmatory for choriocarcinoma, in which blood spaces were surrounded by cytotrophoblast cells that in turn were surrounded by syncytiotrophoblast cells. Also, abnormal mitoses with nuclear pleomorphism and hyperchromasia were noted within the cells due to malignancy (Figure 12). No chorionic villi were seen in the specimen. The tumor was positive for β-HCG on immunohistochemistry. The patient completed a three-month course of chemotherapy and is currently symptom-free with β-HCG levels within normal limits.

Discussion

Ovarian choriocarcinoma is an extremely rare subtype of germ cell tumor, first described in 2004 [1]. Less than 1% of ovarian tumors are choriocarcinomas. Only around thirty
cases have been described worldwide to date. Pure primary ovarian choriocarcinoma can arise from old ovarian ectopic pregnancy (gestational) or de novo without any antecedent inciting cause (non-gestational) [2]. Gestational choriocarcinoma can be distinguished from the non-gestational type by DNA polymorphism analysis, whereby presence of paternal antigens in the specimen suggests the gestational type [3]. However, it is a highly expensive test and could not be performed in our department. On ultrasound imaging, choriocarcinomas appear as solid adnexal masses with cystic areas containing dense mobile echoes and septations due to hemorrhage. Intense vascularity with low-resistance arterial waveforms can be seen on color Doppler. On CT, choriocarcinoma appears as a well-defined, lobulated, hypodense mass with areas of hemorrhage. Intense enhancement is seen on post-contrast images. Prominent ovarian and uterine vessels are also seen, signifying the vascular nature of choriocarcinoma. MRI, with its excellent soft tissue resolution, shows these masses as iso-to-hyperintense lesions on T1, T2, and STIR images, with avid enhancement of the solid portion of the tumor. Areas of hemorrhage appear as hyperintensities on T1W sequences and as blooming on GRE images. Gross surgical specimens
Figure 10. Axial fat-saturated post-contrast T1W images show avid enhancement of both ovarian lesions.

Figure 11. Gross surgical specimen shows bulky ovaries with cystic and hemorrhagic spaces (arrow).

Figure 12. High-power microscopic histopathological specimen shows syncytiotrophoblast and cytotrophoblast cells with nuclear pleomorphism and increased mitoses.

appear as solid masses with blood-filled spaces. The presence of trophoblastic cells and blood spaces with cellular atypia and nuclear polymorphism is specific for choriocarcinoma on the microscopic examination. Absence of intratumor trophoblastic tissue cannot aid in the diagnosis of the non-gestational subtype of primary ovarian choriocarcinoma. Although both gestational and non-gestational types secrete excess β-HCG, the levels of β-HCG are relatively lower in the non-gestational type [4].

The main differential diagnoses of ovarian choriocarcinoma are other germ cell tumors of the ovary, ectopic pregnancy and tubo-ovarian masses due to chronic infection [5]. Treatment of ovarian choriocarcinoma is either chemotherapy, surgery, or a combination of the two [6,7]. However, treatment is dependent on the subtype of chorionicarcinoma [8]. The gestational subtype has been shown to respond better to chemotherapy [9], particularly to bleomycin, etoposide, and cisplatin [10,11]. Cases treated successfully with chemotherapy show reduction in lesion size and β-HCG levels.

Conclusions

Based on clinical history, imaging findings, and serological and histopathological evidence, a final diagnosis of bilateral primary non-gestational choriocarcinoma was made, and the patient was managed accordingly.

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