Case Report

Steroid Cell Tumor of the Ovary – A Rare Case Report and Review of Literature

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Abstract

Steroid cell tumors of the ovary are extremely rare, accounting for only 0.1% of all ovarian tumors. Most steroid cell tumors secrete steroid hormones, and only about 10%–15% of patients are asymptomatic. The clinical presentation may take many forms, including abdominal pain, distention, irregular menstrual cycles, and hirsutism. Here, we present a case of a 60-year-old postmenopausal patient who presented with complaints of bleeding per vagina and abdominal pain for 4 months. Ultrasonography (USG) revealed a hypoechoic left adnexal mass measuring 65 mm × 40 mm × 30 mm. Based on these USG findings, the diagnosis of cystic lesion of the left ovary was made. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done, and the specimen was sent for histopathological analysis. On histopathology, diagnosis of steroid cell tumor-not otherwise specified was offered. This case is reported due to its rarity and its unusual presentation, together with a brief review of the literature of the same.

Keywords: *Left adnexal mass, ovary, steroid cell tumor*

Introduction

Steroid cell tumors of the ovary are rarely identified and constitute only about 0.1% of ovarian tumors. These tumors have been divided into three subtypes according to their cells of origin as follows: stromal luteoma, Leydig cell tumor, and steroid cell tumor-not otherwise specified (NOS).[1,2] Of these subtypes, the steroid cell tumors-NOS account for about 56% of steroid cell tumors.[2]

Most steroid cell tumors are associated with secretion of steroid hormones, which causes symptoms that lead to a clinical diagnosis. In general, testosterone secretion causes virilization or hirsutism; estrogen secretion causes bloating, fibrocystic lumps in the breast, and only 10%–15% of patients have no clinical signs or symptoms of increased hormone levels.[3] Morphologically, steroid cell tumor-NOS presents as solid, well-circumscribed mass, yellow in about 89% of cases. Only rarely, in about 1.6% of cases, these tumors are completely cystic.[2]

The following case is of a steroid cell tumor (NOS) of ovary diagnosed in a 60 year old postmenopausal female. Significant and unusual findings, in this case, are short duration of complaints, the age of the patient, lack of any overt androgenic manifestations, and discrepancies between clinical, radiological, and pathological findings.

Case Report

A 60-year-old postmenopausal female presented with complaints of bleeding per vagina and lower abdominal pain for 4 months. She had four children, and there was no history of any complications during her pregnancy. There was no history of use of any contraception or exogenous hormone intake.

Her past medical and surgical history was unremarkable. No significant findings were noted in her family. Ultrasonography (USG) of the pelvis was done in another hospital which revealed a hypoechoic left adnexal mass measuring 65 mm × 40 mm × 30 mm. Based on these USG findings, the diagnosis of cystic lesion of the left ovary was made by the concerned radiologist, and unfortunately, the radiological image of the same was unavailable. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done, and the specimen was sent to our department for histopathological examination.

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We received a gross specimen of uterocervix with attached right adnexae, uterocervix measuring 8.5 cm × 5 cm × 2.5 cm in size. The cut section of uterocervix was unremarkable. The left ovarian mass was received separately and measured 6.5 cm × 4 cm × 3 cm. The cut surface of the left ovary showed solid, yellow mass replacing the whole ovary without any peripheral ovarian stroma [Figure 1]. Other side ovary was grossly normal.

Sections from the left ovarian mass showed well-circumscribed tumor tissue arranged in lobules, nests, and cords. Individual tumor cells were large, round-to-polygonal showing mild anisonucleosis with round-to-oval centrally placed nuclei with prominent nucleoli and abundant amount of eosinophilic cytoplasm. Occasional mitotic figures were also seen. There was no atypia or necrosis noted in the tumor. Histological features were consistent with steroid cell tumor-NOS type [Figure 2]. The cervix showed features of chronic cervicitis with squamous metaplasia and nabothian cyst, whereas the endometrium showed atrophic changes. The other side ovary and tube were unremarkable.

Discussion

Scully was the first person who described ovarian steroid cell tumor, and he reported 63 cases ranging from 2 to 80 years of age.[1] Formerly, these tumors were referred as lipid or lipoid cell tumors of the ovary. Sex cord-stromal tumors develop from the sex cord and stromal components of the gonads.

The incidence of steroid cell tumors is <0.1% of all the ovarian tumors. These are more common in the child-bearing age group, and also during the third and fourth decades, but very rarely, it can also occur in postmenopausal women. These tumors secrete hormones such as androstenedione, α-hydroxyprogesterone, and testosterone and these cause androgenic manifestations.[2,3]

Steroid cell tumors generally present with symptoms of virilization, particularly hirsutism, menstrual irregularity, abnormal hair growth on the face and legs, and sometimes presents with male-type baldness. Hence, in cases where there is unexplained hirsutism, ovarian and adrenal tumor association should be suspected clinically, and thorough evaluation should be done to rule out any occult malignancies.[4] In many cases, the diagnosis is usually made postoperatively on finding a tumor in the ovary accidentally where these tumors do not show any symptoms of virilization.[5]

A majority of steroid cell tumor-NOS are generally unilateral, benign, and well-circumscribed. The size varies from 1.2 to 45 cm.[1] Grossly, these tumors are commonly solid; however, a combination of solid and cystic form or predominantly cystic form may also be seen. The color of the cut surface may range from yellow to orange to red or brown depending on the lipid content. Areas of hemorrhage and necrosis may also be seen.[1,6]

The tumor in our case was completely solid with no cystic area. The cut surface was typically yellow and lobulated.

Steroid cell tumor, NOS should be differentiated from other steroid cell tumors such as stromal luteoma and Leydig cell tumor. Stromal luteoma is most commonly located in the ovarian stroma, and it frequently occurs in association with stromal hyperthecosis. Another feature which helps in diagnosing this tumor is the presence of degenerative pseudovascular spaces containing red blood cells.[6,7]

Leydig cell tumor is usually present in the hilar location. The tumor cells show cytoplasmic Reinke crystals, and it is usually associated with Leydig's cell hyperplasia.[8]

Pregnancy luteoma may sometimes microscopically resemble steroid cell tumor; it is most commonly multifocal and occurs bilaterally in approximately one-third of the patients. It usually regresses after the pregnancy.[7,9]

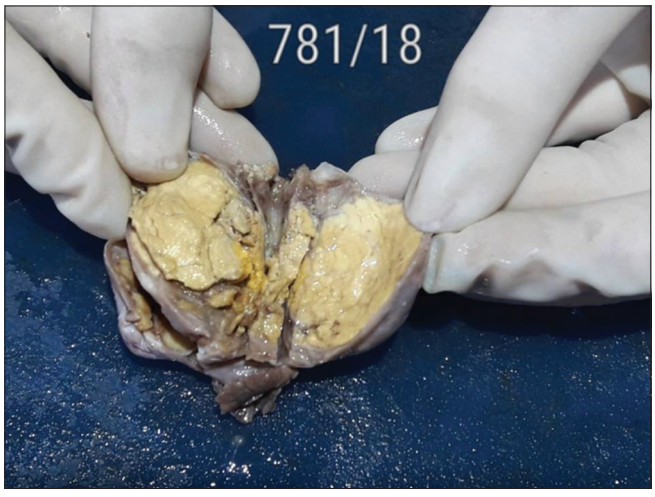

Figure 1: Gross appearance - Cut surface of the left ovary showing solid, yellowish appearance

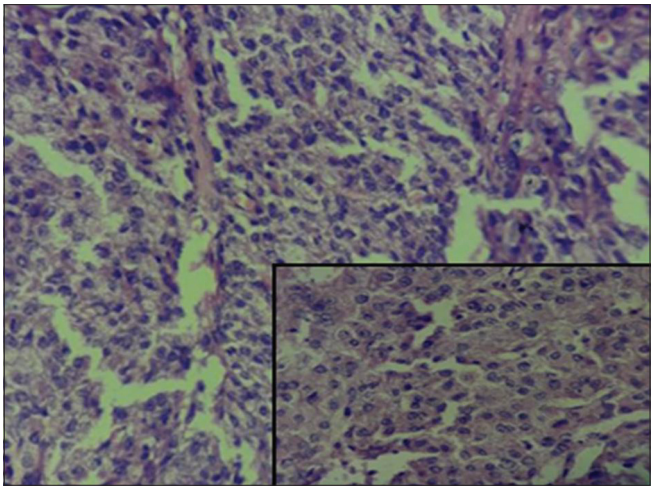

Figure 2: Large polyhedral cells with round-to-oval vesicular nucleus and prominent nucleoli. Cytoplasm is abundant clear and eosinophilic (H and E, ×10 and × 40)

In the present case, features such as stromal hyperthecosis, pseudovascular spaces, or Reinke crystals were absent to suggest the possibility of stromal luteoma and Leydig's cell tumor, and hence, this case was concluded as steroid cell tumor-NOS. Immunohistochemical markers, inhibin and calretinin are quite useful in differentiating this tumor from other non sex cord tumors.[6,8]

A clinicopathological correlation is very important in these tumors as the benign-looking tumors on histomorphology can behave in a clinical malignant manner. The treatment of these tumors should be based on the histological picture, surgical staging, and the patient's desire to preserve fertility. In a young patient who want to preserve their fertility, unilateral salpingo-oophorectomy is the preferred method of treatment.[7]

There are certain clinicopathologic parameters which correlate with adverse behavior of the tumor such as older age at the time of presentation, size of the tumor more than 7.0 cm, mitosis more than 2/10 HPFs, grade 2–3 nuclear atypia, necrosis, and hemorrhage.[5]

In the present case, even though the patient is of older age group, she had a good prognosis as the size of the tumor was <7 cm, and on microscopy, there were occasional mitotic figures without necrosis.

The patient was not offered any further medications or chemotherapy and was advised only follow-up as she had already undergone hysterectomy with bilateral salphingo-oophorectomy. The patient is doing well presently.

Conclusion

Steroid cell tumors-NOS are very rare ovarian sex cord-stromal tumors which are usually associated with various symptoms such as pain in the abdomen, hirsutism, and irregular menstrual cycles. In postmenopausal women who are eligible for surgery, a diagnostic and therapeutic bilateral salpingo-oophorectomy is a safe option. Along with clinical correlation, histopathology is the gold standard which can confirm the diagnosis in most of the cases. In atypical cases, immunohistochemistry can be helpful for accurate diagnosis.[8,9]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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