Steroid cell tumor of Ovary: A case report

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ABSTRACT:
Steroid cell tumors, not otherwise specified (NOS), are rare ovarian sex cord stromal tumors with malignant potential. The majority of these tumors produce several steroids, particularly testosterone. We are presenting this case for its rarity.

KEY WORDS: Steroid cell, Tumors.

INTRODUCTION:
Steroid cell tumors of the ovary account for less than 0.1% of all ovarian tumors and these tumors may present at any age in association with interesting presentations related to the hormonal activity and virilizing properties of tumor. The subtype, not otherwise specified, is associated with androgenic changes in approximately one half of patients with this tumor. [1,2] and average age of occurrence of this tumor is 43 years [3].

CASE REPORT:
A 47-year old woman presented with a history of pain abdomen, amenorrhea and abnormal hair growth on the face of four months duration. Physical examination revealed a male pattern of coarser hair distribution in the beard region and arms. Per abdominal examination showed no abnormality. Ultrasonography of the abdomen and pelvis revealed a solid right adnexal mass measuring 5.7 × 5.0 × 3.0 cm. There was no ascites, retroperitoneal lymphadenopathy, adrenal gland enlargement, or liver metastasis. Clinical and diagnostic evaluation revealed a right adnexal mass and elevated serum levels of testosterone. The patient underwent an exploratory laparotomy, intra-operatively the right ovary showed a 5.5x5x2.5 cm firm, tan-brown, well-encapsulated mass without any adhesions to surrounding structures and right salpingooopherectomy was done. The cut-section of the specimen showed a tan-brown to yellowish orange well-circumscribed tumor Fig: 1,2. Microscopic examination Fig: 3,4 showed diffuse sheets of large polygonal tumor cells with vacuolated
cytoplasm and vesicular nuclei along with prominent nucleoli. Crystals of Reinke, which are usually seen in hilus tumors, were not seen. There was no necrosis or atypical mitosis.

Fig 1: Gross - Smooth surface with intact capsule

Fig 2: Gross - Yellowish orange cut surface

Fig 3: Microscopy - Sheets of Polyhedral cells with eosinophilic granular to clear cytoplasm (low power view)

Fig 4: Microscopy - Pale eosinophilic cytoplasm & small round nucleus (high power view)
DISCUSSION:

Ovarian steroid cell tumours were first described by Scully in 1979[4]. Steroid cell tumours are now included in the World Health Organisation classification and account for approximately 0.1% of all ovarian neoplasms. Ovarian steroid cell tumours comprise three groups: stromal luteomas, Leydig cell tumours (hilus cell tumours, and Leydig cell tumours non-hilar type), and steroid cell tumours not otherwise specified (NOS). The last group accounts for about 60% of steroid cell tumors[5,6,7]. They are grouped under sex-chord stromal tumors and are usually benign, unilateral and characterized by a steroid cell proliferation [1,6]. This tumors can occur at any age, but usually develop in adults with an average age of 43 years. The major symptoms detected in 56-77% of patients are hirsutism and virilisation[8,9]. Reedy et al. reported a case of an undifferentiated NOS steroid cell tumor with hirsutism, amenorrhea, clitoromegaly, and temporal baldness[10]. Steroid cell tumors are associated with androgenic changes with variable frequency, ranging from 12% to 50 %[1,7,11,12].

Around 40% of steroid cell tumors are known to be malignant [3,13]. Histological criteria for malignancy are severe nuclear atypia with two or more mitoses/HPF, necrosis, hemorrhage and tumor diameter >7 cm. Metastasis, however, is the only definite sign of malignancy [14]. On histopathological examination cells are most typically arranged diffusely, but may grow in large nodules or nests, or in cords or columns. A minor fibromatous component and areas of hyalinization are occasionally present. The more common type of tumor cell is polygonal and of medium to large size, with slightly granular, eosinophilic cytoplasm. A second cell type is larger with abundant spongy cytoplasm; transitions between the two cell types are usual. Cytoplasm is usually abundant but may be less copious, particularly in poorly differentiated tumors. The tumor cells typically have distinct cell membranes and central nucleoli with a prominent nucleolus. Intracytoplasmic lipochrome pigment is present in approximately one third of the cases. Nuclear atypia is generally slight or absent, but it is moderate or marked in approximately 25% of cases. The mitotic rate varies and does not clearly correlate with nuclear atypia. Approximately two thirds of tumors with severe nuclear atypia and 80% of those with two or more mitotic figures per 10 high-power fields are malignant [6].

The mainstay of ovarian steroid cell tumor treatment is surgery. Surgical treatments using total abdominal hysterectomy, bilateral salpingo-oophorectomy, and complete surgical staging are an appropriate management option for old women who do not want to preserve their fertility, as was our case. However, in young patients, unilateral salpingo oophorectomy is adequate most of the time due to the low bilateral frequency of 6%. However such practices require a mandatory follow-up evaluation and should include a measurement of sex hormone levels, particularly for those patients who demonstrated elevated levels before removal of the primary tumor. Additionally, a gonadotropin releasing hormone agonist could be used as postoperative adjuvant therapy [15,16].

CONCLUSION

Ovarian steroid cell tumors, grouped under sex-chord stromal tumors, account for less than 0.1% of all ovarian tumors. They are usually benign, unilateral and are characterized by long history of often many years of androgenic changes with variable frequency. The primary treatment is surgical extirpation of the primary lesion, and there are no reports of effective treatment with radiation or chemotherapy.
REFERENCES


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