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Case report

Steroid cell tumor of ovary manifesting with hair loss – A rare presentation

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A R T I C L E I N F O

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A B S T R A C T

Steroid cell tumor is a rare neoplasm which accounts for less than 0.1% of all ovarian neoplasms. Steroid cell tumor, Not Otherwise Specified (NOS), accounts for approximately 60% of steroid cell tumors. These tumors can produce steroids, especially testosterone and may give rise to virilizing symptoms such as hirsutism, hair loss, amenorrhea or oligomenorrhea. We present a case of 29 year old female patient with benign steroid cell tumor who presented with irregular cycles and hair loss and was treated with unilateral salphingo-oophorectomy.

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Introduction

Steroid cell tumors (SCT) are rare sex cord-stromal tumors of the ovary which account for less than 0.1% of all ovarian neoplasms [1]. These tumors were previously classified as "lipid" or "lipoid" cell tumors. The term "steroid cell tumors" was coined by Scully in 1979 [2]. Steroid cell tumors are sub-classified as (1) stromal luteoma arising from ovarian stroma (2) leydig cell tumors (hilus cell tumor and leydig cell tumor, non-hilar type) arising from leydig cell (3) steroid cell tumors, Not Otherwise Specified (NOS) when the lineage of tumor is unknown. We report a 29 year old lady with benign steroid cell tumor presenting with hair loss and menstrual irregularity.

Case report

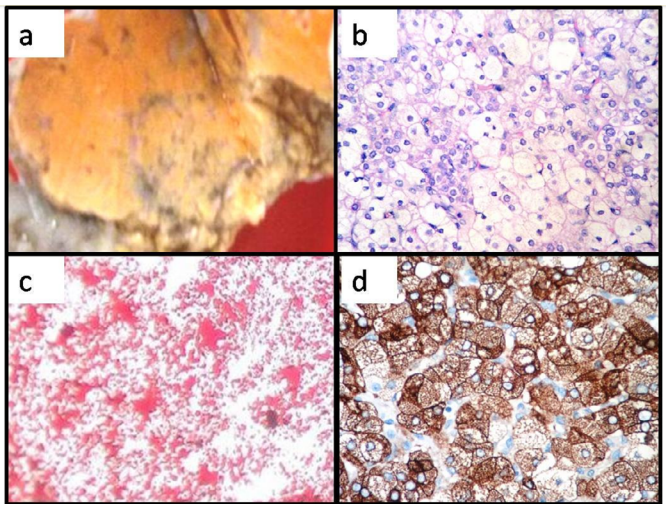
A 29 year old lady; P1L1 with previous caesarean section and a known case of hypothyroidism and hypertension presented with irregular cycles and hair fall since one year. No other virilizing symptoms were present. On examination she was found to have left fornicial mass. USG abdomen showed a left ovarian mass measuring 5x4 cm. Right ovary was normal. The hormonal levels were CA-125- 92 U/ ml , AFP<5 IU/ml , HCG - 6 MIU and LDH- 400 U/L. Laprotomy and left salphingo- oophorectomy was done.

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On gross examination the ovary was measuring 5x3x2 cm. Cut surface showed a well circumscribed nodular, bright orange coloured, solid mass with a rim of normal ovarian parenchyma (Figure 1a). Microscopic examination showed lobules of large polygonal cells separated by fibrovascular septae. The cells had centrally placed uniform small round nucleus with abundant foamy cytoplasm. Mitosis was less than 2/10 HPF. There were no areas of haemorrhage or necrosis (Figure 1b). Oil Red O was strongly positive (Figure 1c) and PAS was negative in the cells indicating presence of lipid. Inhibin was strongly positive (Figure 1d) confirming sex-cord stromal origin of tumor. The patient at present is on follow up and doing well.



Discussion

Steroid cell tumors, NOS is a rare tumor with only few cases reported in literature [3-5]. This tumor is seen in women of reproductive age, particularly during the third and fourth decades, and rarely in postmenopausal women or children. They are clinically androgenic in 40%

of cases and regularly secrete androstenedione, α-hydroxyprogesterone, and testosterone. Virilizing symptoms are seen in 56-77%, estrogenic manifestations in 6-23% , cushing's syndrome in 6-10% and no endocrine symptoms in 25% of the patients [6,7]. The common virilizing symptoms are hirsutism, amenorrhea or oligomenorrhea. Hair loss as seen in this patient is a very rare symptom of virilization.

Steroid cell tumors are usually well circumscribed, unilateral, and solid with cystification due to degeneration and hemorrhage. They vary greatly in size from 0.5 to 45 cm in diameter. The cut surface, which bulges in the fresh state, is frequently lobulated and ranges in color from bright yellow through red-brown to dark green-brown. Microscopically, the Steroid cell tumors consist of rounded or polygonal vacuolated cells arranged in nests or columns and separated by a rich network of capillaries and vascular sinusoids. The tumor cells can show varying degrees of nuclear pleomorphism and mitotic activity, but a definitive diagnosis of malignancy can only really be made by the presence of local invasion. Hayes and Scully have identified five pathologic features that were highly associated with malignancy. They are [1] More than two mitoses per high-power field [2] necrosis [3] size of 7 cm or larger [4] hemorrhage and [5] Grade 2 or 3 nuclear atypia.[2] This patient had the classical microscopic picture of steroid cell tumor with no features suggestive of malignancy.

On immunohistochemmistry majority of the SCT are positive for calretinin and inhibin . Seventy five percent of cases are vimentin positive. Cytokeratin, CD99 and S 100 show variable positivity [8]. This tumor showed strong positivity for inhibin. Other markers were not done due to its classic appearance.

The differential diagnosis for SCT NOS includes [1] Non-neoplastic lesions like Pregnancy luteomas that are more commonly multiple (approximately 50%) and bilateral (approximately 30%). [2] Primary neoplasms like Stromal luteoma which is confined to the ovarian stroma and associated with stromal hyperthecosis. Leydig cell tumors that are localized to the ovarian hilus contain Reinke crystals. Lipid-rich Sertoli cell tumor that have the presence of more typical areas of Sertoli cell tumor, Leutinized granulosa cell tumor or thecomas which have classic areas representative of primary tumor.

Also included are oxyphilic variants of other ovarian tumors (eg, oxyphilic clear cell carcinomas, oxyphilic endometrioid carcinomas), and [3] Metastatic neoplasms like metastatic malignant melanoma, metastatic renal cell carcinoma, and metastatic hepatocellular carcinoma. These can be differentiated from SCT by immunohistochemical features characteristic of these tumors. (eg, Cytokeratin 7 and EMA in clear cell and endometrioid carcinoma; S100 protein and other melanocytic markers in melanoma; CD10 in renal cell carcinoma, and Hepar1 in hepatocellular carcinoma).

The majority of SCTs have a benign or low-grade behavior; and about 25%–45% behave clinically malignant with 20% of cases having an extra ovarian spread of tumor at the time of operation. And 20% of metastatic lesions usually occur within the peritoneal cavity and rarely occur at distant sites. In a series of 63 cases by Scully et al. 28.6% cases had features suggestive of malignant nature[9]. Our patient had benign tumor with no features favoring malignancy clinically and on histopathology.
Treatment for benign SCTs is primarily surgical removal of the tumor. In young patients unilateral salpingo oophorectomy is preferred. In older females total abdominal hysterectomy with bilateral salpingo oophorectomy is done. Malignant tumors require debulking surgery, with post operative chemotherapy or radiotherapy [10]. Our patient was treated with unilateral salphingo- oophorectomy with follow up.

Conclusion

Steroid cell tumor, NOS is a rare functioning ovarian neoplasm with malignant potential. Pathological examination in evaluating the features of malignancy is crucial as benign tumors are cured by salpingo-oophoectomy. Hair loss as seen in this patient is a rare presentation which should raise the suspicion to evaluate and rule out a possible ovarian neoplasm.

Sources of support : nil
Conflicts of interest : nil

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