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Ovarian Steroid Cell Tumor in a 3 year old Female: A Case Report

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Abstract
Steroid cell tumors [SCT] are particularly rare ovarian sex cord-stromal tumors which comprise less than 0.1% of all ovarian tumors. Theses tumors are uncommon in patients prior to puberty without atypical syndromes. We herein present a case of 3 year old female patient who presented with early onset of thelarche, pubic and axillary hair since 6 months and early onset of menarche since 2 months. USG and CECT abdomen and pelvis done which is suggestive of large well defined solid-cystic abdomino-pelvic lesion [5.4 × 6.6 × 7.7cm] noted in right adnexa. Patient underwent laparoscopic assisted right ovarian mass excision and the histopathological examination revealed an ovarian steroid cell tumor and/or leydig cell tumor. Immunohistochemistry confirmed it to be steroid cell tumor. On follow-up patient showed improvement with regression of symptoms.
Keywords: *Ovarian tumor, Steroid cell tumor, Thelarche, Menarche.*

Introduction

Ovarian steroid cell tumors are rare sex cord-stromal tumor of ovary and comprise less than 0.1% of all ovarian tumor[1] .Based on cell of origin they may be divided into 3 subtypes: leydig cell tumor arising from leydig cell in the hilum of ovary, stromal leutoma arising from ovarian stromal cells and steroid cell tumors Not Otherwise Specified (NOS). Last subtype makes up ~ 2/3rd of SCTs and tends to affect younger women (mean age 43 years). SCT NOS are usually benign, however clinically malignant behavior such as peritoneal metastasis occurs in 25-40% of the cases[2,3]. Here we are reporting a case of ovarian steroid cell tumor in a 3 year old female child who presented with early onset of thelarche, pubic and axillary hair since 6 months and early onset of menarche since 2 months. We are going to discuss about presentation, diagnosis and treatment of SCT.

Case Report

A 3 year old female child who presented with early onset of thelarche, pubic and axillary hair since 6 months and early onset of menarche since 2 months. USG and CECT abdomen and pelvis done which is suggestive of large well defined solid-cystic abdomino-pelvic lesion [5.4 × 6.6 × 7.7cm] noted in right adnexa. No evidence of calcification within. On laboratory analysis serum beta HCG < 1.2 mIU/ml (normal range 0-5.3), serum alpha fetoprotein - 3.26IU/ml (normal range 0-5.5), serum

cortisol - 10.78µg/dl (normal range 3-21), LH (Leutinising Hormone) - 1.50 mIU/ml, FSH (Follicle Stimulating Hormone) - 0.46 mIU/ml. RFT, LFT and serum electrolyte were within normal range. Patient underwent laparoscopic assisted right ovarian mass excision. The histopathological report revealed ovarian SCT and/or leydig cell tumor. Immunohistochemistry for confirmation and typing was done, which confirmed it to be Steroid cell tumor.

Gross appearance: tumor measures 11×7×5cm, external surface is smooth shiny and congested. (Fig 1)

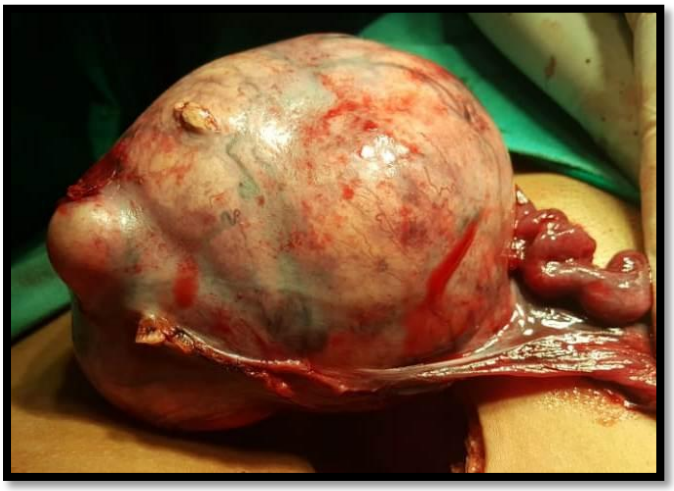

Fig-1 Gross appearance.

On microscopy: multiple section studied shows a well circumscribed and well encapsulated tumor arranged in lobules separated by thin fibrovascular septae. Individual tumor cells are large, polygonal with spongy, granular to clear to eosinophilic cytoplasm with central round to oval nuclei with inconspicuous nucleoli. Mild nuclear atypia and occasional mitosis seen. Reinke crystals and lipochrome pigment are not evident. (Fig-2)

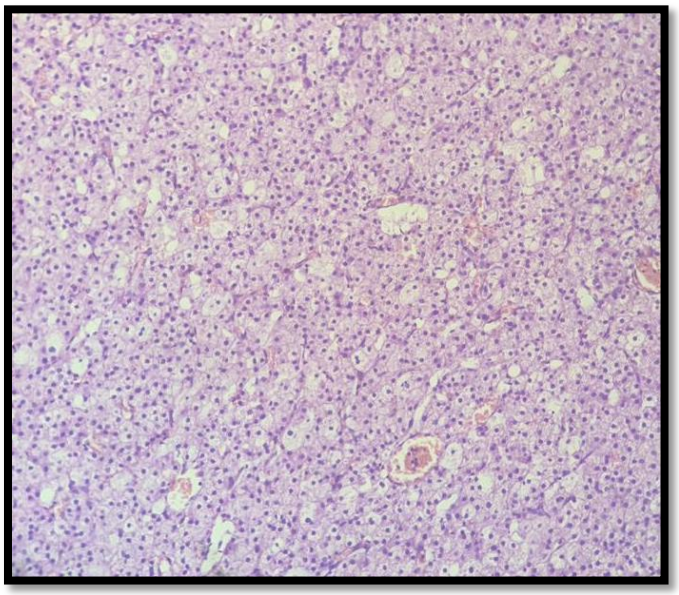

Fig-2 (H & E Stain slide, 20X)

On Immunohistochemistry: tumor cells are positive for mic2, Inhibin and weakly positive for calretinin while negative for AE1/ AE3.

Discussion

The term Steroid cell tumor not otherwise specified was first used by Scully and signifies that the cell lineage is not defined; thus they cannot be categorized as either stromal leutomas or leydig cell tumors[4,5] .Approximately 56-77% of the cases are clinically associated with androgenic changes, such as hirsutism and virilization; 6-7% of the cases are clinically associated with Cushings syndrome; and 25% of SCT-NOS are non-functional. Ovarian SCT-NOS may occur at any age (mean age 43 years) and occasionally before puberty[6] .In our case the patient was aged 3 years without any changes in blood pressure, virilization or hirsutism.

According to the study of Hayes and Scully, the following pathological characteristics may indicate malignancy: ≥ 2 mitotic figures/10 high power fields is associated with 92% risk of malignancy; necrosis with an 86% risk of malignancy; a diameter >7cm with 78% risk of malignancy; hemorrhage with 77% risk of malignancy; and grade 2 or 3 nuclear atypia with a 64% risk of malignancy[6]. To date, 24 cases of ovarian SCT in young female patients aged 2.5-13 years have been reported, but none have been malignant[6-10].

Therefore, ovarian masses suspicious for SCT in children at an early stage should be approached conservatively, unless distinct signs of metastasis are present at the time of presentation[11]. Immunohistochemistry is particularly useful for the accurate diagnosis of SCT. Moreover, unilateral salpingo-oophorectomy is generally considered to be adequate for pediatric patients with stage Ia disease[6].

Conclusion

Early diagnosis of ovarian tumors is important for the prognosis of patient. In this case early diagnosis lead to early management and surgical intervention which prevented complication.

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