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*Case Report*

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# Ovarian steroid cell tumor, not otherwise specific, causes Cushing syndrome in 6 years old girl: A case report

***Versha Rani Rai; Karishma Rahak*; Maira Riaz; Shazia Mahar; Ikramullah Shaikh; Shafqat; Mohsina Noor Ibrahim***

*National Institute of Child Health, Karachi, Pakistan.*

***Corresponding Author: Kirishma Rahak**

National Institute of Child Health, Karachi, Pakistan.

Email: dr.rahak@yahoo.com

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## Abstract

We report a case of 6 years old girl with ovarian Steroid Cell Tumor, Not Otherwise Specific (SCT-NOS). She was admitted to the hospital with increased weight gain, hypertrichosis, and cushingoid facies in form of prominent cheecks, moon facies with dorsocervical fat pad, obese built, acanthosis nigricans on back of neck and marked hirsutism.

Initial workup showed raised level if cortisol with loss of diurnal rhythm, and undetectable Plasma ACTH levels, increased testosterone levels. Ultrasound revealed left sided adnexal mass having no flow inside, which was later confirmed on CT as neoplastic lesion of left ovary measuring 6.9 X 8.1 X 4.9 cm. The patient underwent left oophorectomy and pathological examination showed steroid cell tumour, limited to ovary and negative for malignancy. She was kept on IV hydrocortisone preop and post operatively and antihypertensives. After tumor resection her testosterone and cortisol returned to normal levels.

***Keywords:*** steroid cell tumor; not otherwise specific; cushing syndrome; ovarian mass.

## Introduction

SCT-NOS in children and adolescents are extremely rare, with only about 9 cases found in a PubMed search, and first case reported in our country. In some pediatric SCT-NOS cases, the functional features of steroid hormone-secreting cells produce characteristic clinical signs of virilization and Cushing's syndrome. The majority of these tumors have a benign character, but a small portion of them behave in a malignant manner. In this report we describe the case of a 6 year old girl presented with increased weight gain, hirsutism, cushingoidfacies. SCT-NOS of the ovary should be considered in cases of childhood virilization. In addition, treatment for this tumor type and follow-up regarding endocrinological symptoms are further explained in the discussion section.

## Case report

A 6 year old female patient admitted with complains of weight gain and hirsutism for past 10 months. On examination she had obvious cushingoid facies (Figure 1A), facial hair growth, hypertrichosis (Figure 1B), dorsocervical fat pad, acanthosis nigracans and marked hirsutism with ferriman Gallwey score of 25.

Her blood pressure was >99$^{th}$ centile whereas tanner staging was P3 A1 B1.

Abdominal ultrasound detected left sided adnexal mass having no flow inside. On CT abdomen it was a left adnexal mass of 6.9 X 8.1 X 4.9 cm (CC X TS X AP measurements. Investigation showed increased level of morning cortisol 28.10 ug/dl with loss

of diurnal rhythm and undetected plasma ACTH. Testosterone level 118.8 ng/dl which was four times normal. Thyroid profile, FSH, LH, beta. HCG, alpha fetoprotein all were in normal ranges.

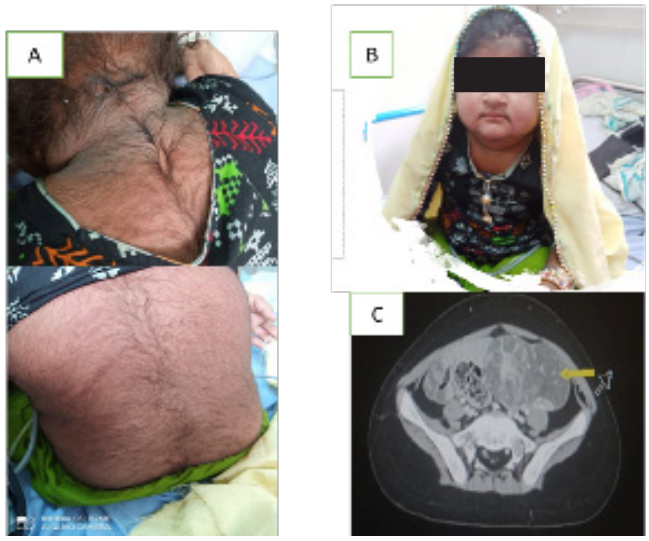


**Figure 1:** Clinical amd imaging findings of steroid cell tumour patient. Photographhs shown hirsutism of neck and back (A). Moon face, prominent cheeks and hirsutism (B). CT Aial image shows heterogenous lesion in left adenexa (C).

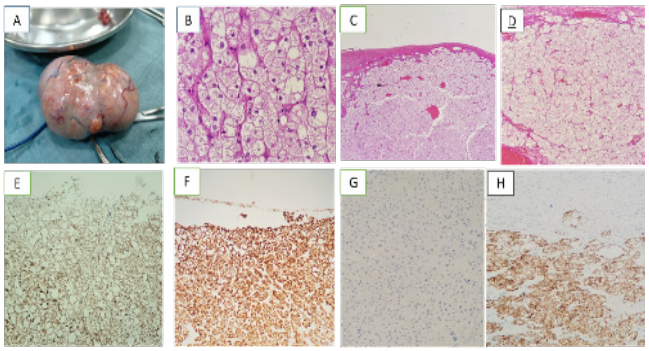


**Figure 2:** Gross appearance and pathologic findinds of steroid cell tumour. Smooth tumour surface (A). Polygonal cells with distint membrane and abundant esinophilic cytoplasm (B). Tumor composed of ill defined nodules (C). Tumor composed of trabecular and sheets with intervening fibrous septa (D) immunohistochemistry showing calretinin (E), Inhibin (F) SALL 4 (G). Melanin positive cells(H).

She underwent left oophorectomy. The surface of tumor was smooth without adhesions to surrounding area (Figure 2A). Right ovary and peritonium was normal on intraopertative finding. Mass was present with findings of intact capsule and dilated vessels (Figure 2C). Histopathological report revealed steroid cell tumour NOS (Not Otherwise Specified) which was negative for malignancy. Immunohistochemical strain performed which shows positive inhibin (Figure 2F), calretenin (Figure 2E), Melanin A (Figure 2H) and CD68. She was kept on intravenous hydrocortisone pre and post operative day to avoid adrenal crises. Postoperatively her levels of cortisol and testosterone became normal.

Tab captopril was started for high BP. Postoperatively cortisol and testosterone levels returned to normal level. 4 months after treatment the patient has been without tumor recurrence, in outpatient follow up. Signs of cushing syndrome and hirsutism have improved with no evidence of postoperstive adrenal insufficiency.

## Discussion

Steroid cell tumors of the ovary account for 0.1% of all ovarian tumors and are rarest tumours that cause virilization in children [1].

These tumors are divided into three subtypes according to their cells of origin: stromal luteoma, leydig cell tumor and steroid cell tumor, Not Otherwise Specified (NOS). Among these subtypes, the steroid cell tumors, NOS constitute for about 56% of steroid cell tumors [2]. Commonly, androgenic manifestations in form of virilisation particularly hirsutism are seen in these tumors as they secrete hormones like androstenedione, α hydroxyprogesterone, and testosterone [3]. However there may be features of cushing syndrome as seen in our case due to increased secretion of cortisol. Macroscopically these tumors are yellow orange due to high fat content. The tumor is commonly confined to ovary but rare presentation of extraovarian steroid cell tumor have been found [4]. It is distinguished from leydig cell tumour by absence of reinke crystals in cytoplasm which is hallmark of leydig cell tumor. Definitive diagnosis is made by histology. Mostly these tumors are unilateral an benign, however 25-43% of steroid cell tumors are clinically malignant with 20% cases exhibiting metastasis outside the ovary [5]. A study by hayes and scully [6] documented five pathological features indicative of malignancy as shown in Table below.

| Microscopic features | % chance of malignancy |
|---|---|
| 1) Two or more mitosis per 10 high-power field. | 92 |
| 2) Necrosis | 86 |
| 3) Size of tumor more than 7cm | 78 |
| 4) Hemorrhage | 77 |
| 5) Grade 2 or 3 nuclear atypia | 64 |

In our case, there were no mitotic activity, cytologicatypia, hemorrhage or necrosis, tumor size was 4.1 X 3.5 cm. The treatment depends upon age of patient, clinical features, tumor histology, surgical staging and desire to preserve fertility. In younger age girls conservative surgery with unilateral oophorectomy is performed as in our case because there were no signs of malignant potential. Her harmone levels rapidly declined to normal levels after surgery. Neither relapse nor metastasis was detected posoperatively at 6$^{th}$ month evaluation.

## Conclusion

Steroid cell tumors are rarely found in childhood. This case emphasizes the importance of early consideration of tumor causing cushingoid features and virilization. Early diagnosis is possible with clinical correlation along with histopathlogy of tumor mass in most cases. Definative treatment is with surgical resection of mass and further management is decided on tumor pathology.

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