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

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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 cheeks, 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.
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.

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 postoperative 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.

<table>
<thead>
<tr>
<th>Microscopic features</th>
<th>% chance of malignancy</th>
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<tbody>
<tr>
<td>1) Two or more mitosis per 10 high-power field.</td>
<td>92</td>
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<tr>
<td>2) Necrosis</td>
<td>86</td>
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<tr>
<td>3) Size of tumor more than 7cm</td>
<td>78</td>
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<tr>
<td>4) Hemorrhage</td>
<td>77</td>
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<tr>
<td>5) Grade 2 or 3 nuclear atypia</td>
<td>64</td>
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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 postoperatively at 6th 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 histopathology of tumor mass in most cases. Definitive treatment is with surgical resection of mass and further management is decided on tumor pathology.
References


