Cushing Syndrome in a 6 Years Old Girl due to Adrenal Mass Presented with Seizure

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ABSTRACT

Introduction: Cushing syndrome is due to excessive cortisol level resulting from iatrogenic or endogenous excess production, which may be due to hypersecretion of ACTH from anterior pituitary or pituitary adenoma or by adrenal mass.

Case report: A 6 year old girl presented with seizure, hypertension and features of cushing syndrome with significant virilization including facial hirsutism, excessive pubic hairs, enlarged clitoris and deepening of voice. Investigation revealed a well defined right suprarenal mass on abdominal CT with significantiy high level of serum cortisol, testosterone and 17-α hydroxyprogesterone.

Conclusion: This is a case of cushings syndrome with virilizing adrenal tumor which caused rapid masculinization of previously normal female child.

Keywords: Cushing Syndrome, Adrenal Mass

INTRODUCTION

Cushing syndrome results from abnormally high level of cortisol or other glucocorticoids which may be due to either iatrogenic or excess endogenous secretion.[1] High level of cortisol may result from abnormal secreting adrenal tumor or hypersecretion of ACTH by anterior pituitary or by a tumor.

The most common cause of Cushing syndrome is iatrogenic administration of glucocorticoid hormone or drugs in high doses. Endogenous cushing syndrome is most often due to adrenocortical tumor (adenoma or carcinoma) in infants and young children, whereas in older one (>7yrs.) it is mostly due to excessive ACTH secreting adenoma causing bilateral adrenal hyperplasia.[2] These patients often exhibit signs of hypercortisolism along with features of hypersecretion of androgen, estrogen, and aldosterone. Rare causes of ACTH dependent cushing syndrome in children include ectopic ACTH secreting tumor of Islet cell carcinoma, neuroblastoma, Wilms tumor, hemangiopericytoma, and thymic carcinoid.[3]

Clinically patient presents with mooning of face, generalized obesity, easy bruisability, hypertension and signs of abnormal masculinisation of female child, like hirsutism of face, excess pubic hairs, acne, enlargement of clitoris and deepening of voice. Her growth is impaired and often fall below 3rd percentile except in case where there is significant virilization. Hypertension is common in cushing syndrome and is found more often with ectopic ACTH syndrome, which may cause heart failure. Patient is often susceptible to infection and may lead to sepsis.

CASE REPORT

A 6 year old girl presented at ANMCH, Gaya Bihar, Department of Peditrics with generalized tonic-clonic seizure starting from facial twitch followed by involvement of the whole body and transient loss of consciousness.Seizure lasted for about 5-10 minutes and was not associated with fever, headache, or vomiting. There were three such episodes of seizure within a period of last 6 months. She also developed gradual swelling of whole body with mooning of face, slight obesity, progressive deepening of voice, hirsutism of face and excessive pubic hairs. These all occurred rapidly within a period of six months before that she was completely alright. Her birth history was uneventful and born at term vaginally, cried immediately after birth. She was exclusively breastfed for 6 months, vaccinated properly and all developmental milestones were within normal range. There was no significant illness in the family.

On examination, patient recovered from seizure, become conscious and alert after a brief period of stupor, her vitals were normal except blood pressure which was 180/130 mm Hg. which may be the cause of seizure (hypertensive encephalopathy). Pulse was 86 bpm, regular, fair in volume, no radio femoral delay and all peripheral pulses were palpable. There was no cyanosis, clubbing, lymphadenopathy or pitting edema. There was mooning of face along with mild generalized obesity with short stature (Figure 2), having height 104 cm which is < 3rd percentile for the age and sex, weight 20 kg which was normal for the age and sex. There was marked facial hirsutism (Figure 1) abundant pubic hairs with enlarged clitoris (Figure 3) and mature genitilia of adult type (Figure 4). Blood pressure poorly controlled despite of administration of several drugs like Ca-channel blockers, vasodilators (Labetalol), and ACE-Inhibitor. Other systems like CVS, CNS, and abdomen showed no significant clinical abnormality.

DISCUSSION

Adrenal tumor are the most common cause of ACTH independent cushing syndrome in young children which may either be due to adrenal adenoma or carcinoma. Adrenal adenoma secretes mostly cortisol with minimal secretion of mineralocorticoid or sex steroid whereas carcinoma tend to secrete excess cortisol, mineralocorticoids as well as androgen.[4] Adrenal carcinoma is more likely to occur in a young female child having early age of onset, rapid progression with features of androgenism, hypertension, and large abdominal mass. Tumor size > 10 cm identified at abdominal USG with vascular invasion of capsule and elevated metabolites like 17 α OH progesterone, androstenedione etc. suggestive of carcinoma.[5] In our case the abnormal laboratory findings include high serum cortisol in the range of 27.04ng/ml with very high serum testeron level

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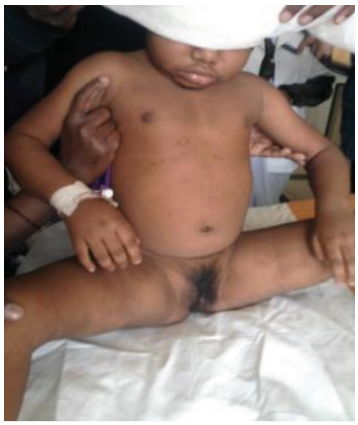

Figure-1: Hirsutism

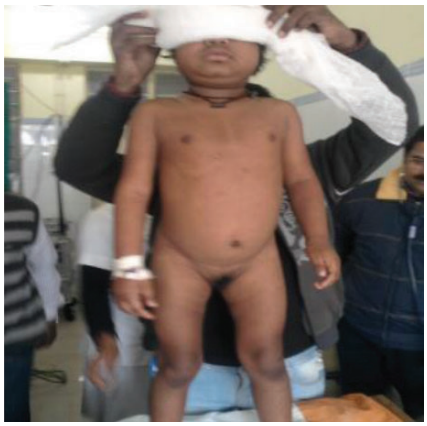

Figure-2: Obesity

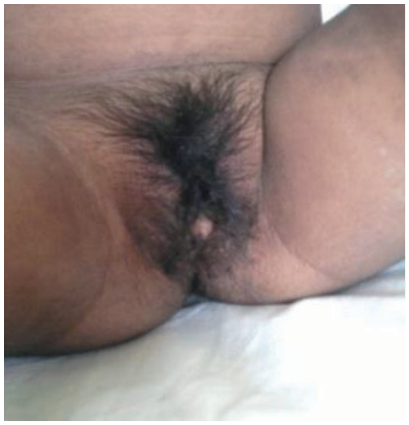

Figure-3: Enlarged clitoris with abundant pubic hairs

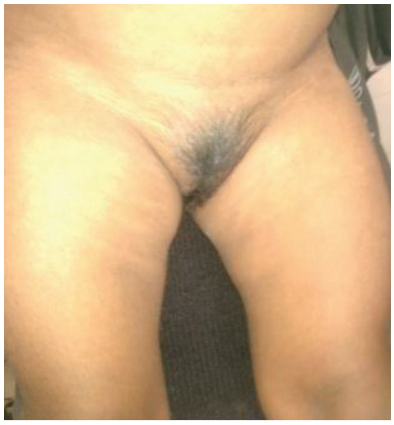

Figure-4: Adult like genitilia

581.62ng/dl and high 17-αhydroxyprogesteron (17-OHP) 7.76ng/ml. Serum ACTH level is 12.5pg/ml. Urinary VMA is 9.31mg/gm of urinary creatinine. These all suggest malignant adrenal tumor as the disease also progressed very rapidly.[7] Abdominal ultrasonography showed 5.2×4.1cm hypo-echoeic mass on the upper pole of right kidney. CT abdomen also suggest well defined mass (4.6×4.6cm) on the right suprarenal region abutting segment VI of liver.

Treatment must be individualized. Lesion 3cm or less should have to be followed up clinically with periodic imaging. Resection of adrenal mass is recommended for adrenal adenoma as well as carcinoma.[6] Transperitonial approach has been recommended but laparoscopic removal may be possible in some cases. Larger tumor >200mm, symptoms >6months, high urinary keto-steroid and 17-OHP have been associated with poor prognosis.[8] Adrenal carcinoma is highly malignant and having high rate of recurrence.

CONCLUSION

In view of above clinical finding and discussion i.e., in a young girl with rapid development of cushingoid habitus with features of virilization and poorly controlled hypertension along with laboratory evidence of high cortisol, increased 17 α- OH progesterone, very high testosterone level, with a heterogenous adrenal mass, we can conclude that this may be a case of adrenal carcinoma causing cushing syndrome, however it should have to be confirmed by biopsy.

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