Case Report

Virilizing lipoadenoma of the adrenal gland in a pre-pubertal girl: A rare case

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ABSTRACT

We report a case of a 12-year-old girl who presented with the history of hirsutism. On evaluation, she was found to have testosterone secreting adrenal gland tumor. Histopathological examination of the adrenalectomy specimen revealed a lipoadenoma.

Key words: Adenolipoma, adrenal gland, hirsutism, virilization

INTRODUCTION

Adrenocortical tumors are rare in childhood and adolescence. Virilizing lipoadenoma of the adrenal gland is an extremely rare tumor. We encountered a patient with this pathology and report the case and review the literature on the subject.

CASE REPORT

A 12-year-old girl presented to our hospital with a history of abnormal menstruation and progressive hirsutism. She had a normal childhood until 2 years back when she started noticing appearance of hair on her face, body, extremities and pubic region. Her height, weight and body mass index were within the normal limits. Blood pressure was 104/64 mm Hg and pulse rate was 92 beats/min. There was thick black hair on the face, arms, chest, back, legs and over the pubic region. Her breasts were pre-pubertal. Pelvic examination showed enlargement of the clitoris. Results of routine lab studies including serum electrolytes were normal.

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Abdominal and pelvic ultrasonography showed the presence of 10 cm × 8 cm sized well- defined, primarily hyperechoic mass lesion seen in the right adrenal region with indentation of the adjacent surface of liver and kidney. The uterus appeared small in size. Multi-detector computed tomography (MDCT) of abdomen, plain and contrast, showed a large well-defined heterogeneously enhancing predominantly fat attenuating lesion measuring 9.5 cm × 8.7 cm × 8.0 cm involving the right adrenal gland. The lesion demonstrated multifocal area of nodular calcification with enhancing solid component and was seen compressing the superolateral aspect of the right kidney with the minimal inferomedial displacement [Figure 1a and b]. Blood tests were carried out for functional assessment of the right adrenal tumor. Serum testosterone level was raised to 2.24 ng/ml. Serum adrenocorticotrophic hormone, 17 hydroxyprogesterone, progesterone and cortisol levels as well as urinary 17 keto steroids, 17 hydroxy corticosteriods and vanilylmandelic acid levels were normal. A clinical diagnosis of adrenal myelolipoma was made and right adrenalectomy was performed. The specimen showed an encapsulated tumor measuring 10 cm × 8 cm × 7 cm [Figure 1c]. Microscopic examination showed compressed adrenal tissue with foci of hemorrhage within the well- encapsulated tumor. The tumor nodules were primarily composed of eosinophilic lipid poor cells resembling zona reticularis. Mitotic rate was <5/50 hpf. No atypical mitosis was seen. The tumor nodules were separated by bands of hyalinized fibrous tissue containing proliferating thick walled vessels, hemorrhagic foci and sparse lymphocytic infiltrate. Islands of abundant mature adipose tissue were also seen. Scattered foci containing lipofuscin pigment were seen and no vascular/capsular invasion or necrosis was seen [Figure 1d].

Based on the size, weight of tumor, microscopic features and Weiss criteria a final diagnosis of lipoadenoma, a morphological variant of adenoma was made.

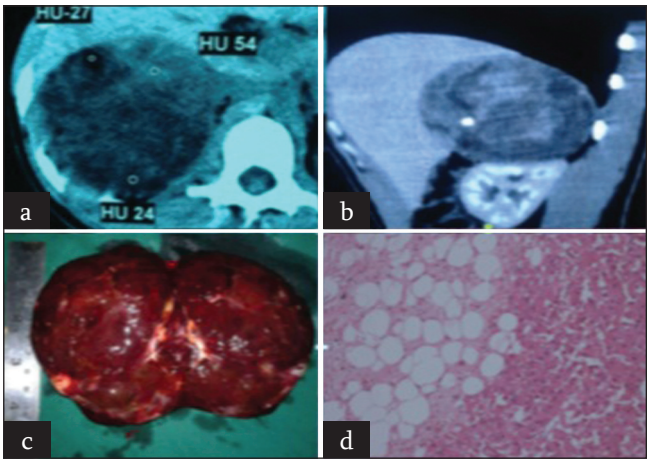

Figure 1: (a) Contrast enhanced computed tomography axial section, showing a 9.5 cm × 8.7 cm × 8.0 cm well-defined heterogeneously enhancing predominantly fat attenuating lesion involving the right adrenal gland. There are multifocal areas of nodular calcification with enhancing solid components. (b) Contrast enhanced computed tomography sagittal section showing the adrenal mass compressing the superolateral aspect of the right kidney with minimal inferomedial displacement. (c) Specimen. (d) Histopathology

Post-operatively serum testosterone level returned to 0.158 ng/ml, which was within the normal limits. Patient's menstrual cycles regularized and features of hirsutism decreased [Figure 2].

DISCUSSION

Adrenocortical tumors are rare in childhood and adolescence.[1] The world-wide annual incidence ranges from 0.3 to 0.38/million children below the age of 15 years with 65% of them occurring in children younger than 5 years of age.[2] The incidence of adrenal cortical tumor seems to be higher in young girls with a female:male ratio of 2:1, whereas in adolescence the female:male ratio is 1:1.[3]

Virilization is the most common presentation in a functioning adrenal gland tumor, followed by hypercortisolism and hyperaldosteronism. In our case, the patient presented with features of hirsuitism and menstrual irregularity. The diagnosis was supported by elevated serum testosterone suggesting the diagnosis of functional virilizing tumor. MDCT showed heterogeneously enhancing predominating fat attenuating lesion suggesting the possibility of myelolipoma.

Mature adipose tissue in adrenal tumors appears in myelolipomas and, in rare cases, lipomas.[4] Rhodes *et al.* described entrapment of adjacent retroperitoneal fat by a hemorrhagic adrenal adenoma.[5] Feldberg *et al.* reported a case of adrenal cortical adenoma with extensive fat cell metaplasia.[6] A number of hypotheses have been put forth regarding histogenesis of lipoid tissue. They are (a) embryonal nests of adipose tissue[7] (b) fatty metaplasia of adrenal gland[8] (c) lipomatous differentiation of previously uncommitted mesenchymal cells within the cortical stroma.[9] In our patient, the adrenal cortical tumor had a large component of adipose tissue dispersed throughout it. As there was no hematopoetic tissue, the possibility of myelolipoma was ruled out. Thus, the tumor was designated as lipoadenoma, one of the morphological variant of benign adenoma of the adrenal gland.

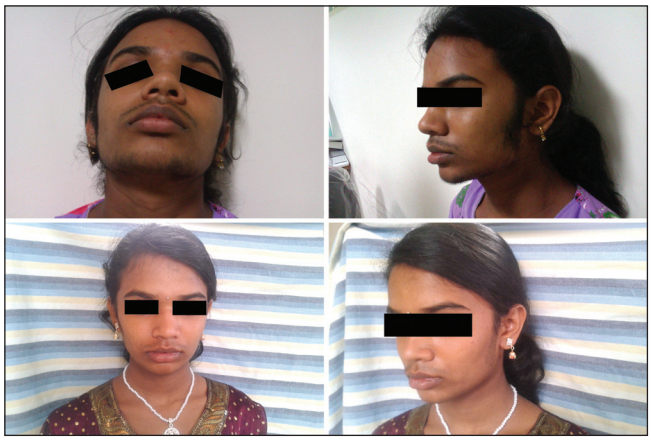

Figure 2: Top 2 images: pre-operative pictures demonstrating thick facial hair. Bottom 2 images: post-operative pictures demonstrating resolution of hirsutism

Assessing the malignant potential of an adrenal tumor is difficult. Weiss criteria are employed to distinguish between benign and malignant tumors. Even the established histopathological criteria and algorithm adapted from tumor in adults generally do not allow the clear classification in children. In the pediatric population if the tumor size is >10 cm, weight >400 g and mitosis >15/20 hpf, it is considered malignant.[10]

In the present case, the size of the tumor was <10 cm, weight of the tumor was 200 g and mitosis was <5/50 hpf. Hence in view of the above features and the histopathological examination, a diagnosis of benign virilizing lipoadenoma of the adrenal gland was made.

CONCLUSION

Adrenocortical tumors in children are extremely rare neoplasm and virilizing lipoadenoma, one of the morphological variant of adrenal adenoma is still rarer and only few isolated cases have been reported in the literature. Virilizing huge adrenocortical tumors in children are viewed with concern for their malignant potential. Good histopathological examination helps in making an accurate diagnosis.

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