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

Prasad Mylarappa, Amey Pathade, Tarun Javali, D. Ramesh
Department of Urology, MS Ramaiah Medical College and Hospital, Bangalore, Karnataka, India

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. 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 adrenocorticotropic hormone, 17 hydroxyprogesterone, progesterone and cortisol levels as well as urinary 17 keto steroids, 17 hydroxy corticosteroids and vanilmandelic 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.
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. 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. 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.

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 hirsutism 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. Rhodes et al. described entrapment of adjacent retroperitoneal fat by a hemorrhagic adrenal adenoma. Feldberg et al. reported a case of adrenal cortical adenoma with extensive fat cell metaplasia. A number of hypotheses have been put forth regarding histogenesis of lipoid tissue. They are (a) embryonal nests of adipose tissue (b) fatty metaplasia of adrenal gland (c) lipomatous differentiation of previously uncommitted mesenchymal cells within the cortical stroma. 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.

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.

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.

**REFERENCES**

1. Michalkiewicz E, Sandrini R, Figueiredo B, Miranda EC, Caran E, Oliveira-Filho AG, et al. Clinical and outcome characteristics of children with adrenocortical tumors: A report from the international pediatric adrenocortical tumor registry. J Clin Oncol 2004;22:838-45.
2. Stiller CA. International variations in the incidence of childhood carcinomas. Cancer Epidemiol Biomarkers Prev 1994;3:305-10.
3. Cagle PT, Hough AJ, Pysker TJ, Page DL, Johnson EH, Kirkland RT, et al. Comparison of adrenal cortical tumors in children and adults. Cancer 1986;57:2235-7.
4. Rosai J. Adrenal gland and other paraganglia. In: Rosai J, editor. Ackerman’s Surgical Pathology. 7th ed. Washington: C. V. Mosby; 1989. p. 789-819.
5. Rhodes RE, Gaede JT, Meyer GA. Hemorrhagic adrenal adenoma simulating myelolipoma: CT evaluation. J Comput Assist Tomogr 1992;16:301-4.
6. Feldberg E, Guy M, Eisenkraft S, Czernobilsky B. Adrenal cortical adenoma with extensive fat cell metaplasia. Pathol Res Pract 1996;192:62-5.
7. Wooley PG. Heteroplastic bone and bone marrow formation associated with tuberculosis in the adrenal. J Lab Clin Med 1915;1:502.
8. Damjanov I, Katz SM, Catalano E, Mason D, Schwartz AB. Myelolipoma in a heterotopic adrenal gland: Light and electron microscopic findings. Cancer 1979;44:1350-6.
9. Boudreaux D, Waisman J, Skinner DG, Low R. Giant adrenal myelolipoma and testicular interstitial cell tumor in a man with congenital 21-hydroxylase deficiency. Am J Surg Pathol 1979;3:109-23.
10. Lau SK, Weiss LM. The Weiss system for evaluating adrenocortical neoplasms: 25 years later. Hum Pathol 2009;40:757-68.

How to cite this article: Mylarappa P, Pathade A, Javali T, Ramesh D. Virilizing lipoadenoma of the adrenal gland in a pre-pubertal girl: A rare case. Indian J Urol 2014;30:219-21.

Source of Support: Nil, Conflict of Interest: None declared.

Author Help: Online submission of the manuscripts

Articles can be submitted online from http://www.journalonweb.com. For online submission, the articles should be prepared in two files (first page file and article file). Images should be submitted separately.

1) First Page File:
Prepare the title page, covering letter, acknowledgement etc. using a word processor program. All information related to your identity should be included here. Use text/rtf/doc/pdf files. Do not zip the files.

2) Article File:
The main text of the article, beginning with the Abstract to References (including tables) should be in this file. Do not include any information (such as acknowledgement, your names in page headers etc.) in this file. Use text/rtf/doc/pdf files. Do not zip the files. Limit the file size to 1024 kb. Do not incorporate images in the file. If file size is large, graphs can be submitted separately as images, without their being incorporated in the article file. This will reduce the size of the file.

3) Images:
Submit good quality color images. Each image should be less than 4096 kb (4 MB) in size. The size of the image can be reduced by decreasing the actual height and width of the images (keep up to about 6 inches and up to about 1800 x 1200 pixels). JPEG is the most suitable file format. The image quality should be good enough to judge the scientific value of the image. For the purpose of printing, always retain a good quality, high resolution image. This high resolution image should be sent to the editorial office at the time of sending a revised article.

4) Legends:
Legends for the figures/images should be included at the end of the article file.