A CASE REPORT OF PRIMARY PIGMENTED NODULAR ADRENOCORTICAL DISEASE

Ho Thi Ngoc Bich - Huynh Thoai Loan - Tran Thi Bich Huyen - Dao trung Hieu

Children's hospital I

57th ESPE 2018 Meeting ATHENS GREECE 27-29 September 2018

ABSTRACT

- Primary pigmented nodular adrenocortical disease (PPNAD) is a rare cause of ACTH-independent Cushing's syndrome and has characteristic gross and microscopic pathologic findings.
- We report a case of PPNAD in a 11-year-old girl. She was admitted to our hospital with a chief complaint of rapid weight gain in 1.5 years. Examination associated with laboratory tests detected hypertension and ACTH- independent cushing syndrome. Images of bilateral nodular adrenal hyperplasia were revealed by abdominal CTScan. Total bilateral adrenalectomy following by renal hormones therapy was the choice of treatment. After 3 months of surgery, improvement of hypertension, weight gain and cushing syndrmome were noted. In addition, there was normalization of adrenal hormones levels.
- Primary pigmented nodular adrenocortical disease (PPNAD) is a rare cause of ACTH-independent Cushing's syndrome. Total bilateral adrenalectomy followed by hormone therapy is the optimal treatment. Assessment of cushing syndrome's improvement associated with periodic evaluation of CNC should be performed.

CONTACT

<Ho Thi Ngoc Bich>

<Children's hospital I>

Email: ngocbichho0587@gmail.com

Phone: +84 0908136873

INTRODUCTION

Primary pigmented nodular adrenocortical disease (PPNAD) and ACTH dependent macronodular adrenal hyperplasia account for 10 percent ACTH independent cushing's syndrome (CS). PPNAD is characterized by pigmented adrenocortical nodules ranging in size from sub-microscopic to 10 mm in diameter. Half of PPNAD appear to be isolated and the other half are familiar, mostly associated with Carney complex (CNC). PPNAD may manifest by typical signs of CS, subclinical or cyclic CS. The optimal choice of treatment for PPNAD-induced CS is bilateral adrenectomy followed by adrenal hormones replacement.

CASE REPORT

A 11 year old girl was admitted to Children's hospital I with the chief complaint of rapid weight gain. This problem had lasted for 1.5 year. In the first year, the body weight increased 10 kg. As a result, obesity was diagnosed by a local hospital and subsequently encouraged to lose weight. This condition continued to aggravate in 6 months before admission to our hospital. In addition, other signs appeared: hirsutism, acne, striae. This condition was not detected in other members of the family.

Clinical findings: hypertension (BP 130 -140/80), hirsutism, round face, striae, short stature (132 cm: 3th percentile for height).

Laboratory tests: midnight salivary cortisol: 12.6 ug/dL (< 0,35 ug/dL) , 24 hour urinary Cortisol: 564.5 ug/mL (Normal < 70 ug/m2/24h), ACTH (morning): <1 pg/mL (10-20 pg/mL), low dose dexamethasone suppression test: cortisol 17 ug/dL (normal< 1,8 ug/dL). Abdominal ultrasound: bilateral nodular adrenal hyperplasia. Abdominal CT scan showed 5 -6 mm diffuse nodules in bilateral adrenal glands.

Treatment: Antihypertensive drugs, bilateral laparoscopic adrenalectomy and adrenal hormone replacement.

Results: After surgery, hypertention improved, antihypertensive drugs were gradually withdrawn. Weight gain condition ceased. After 3 months: No hypertension, weight lost (3 kg/3 months), normal serum cortisol concentration.

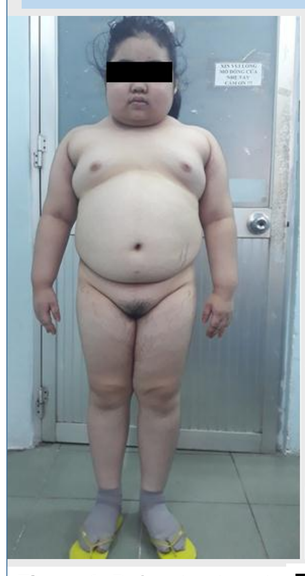

Figure 1. Before treatment

Figure 2. After 3 months of treatment

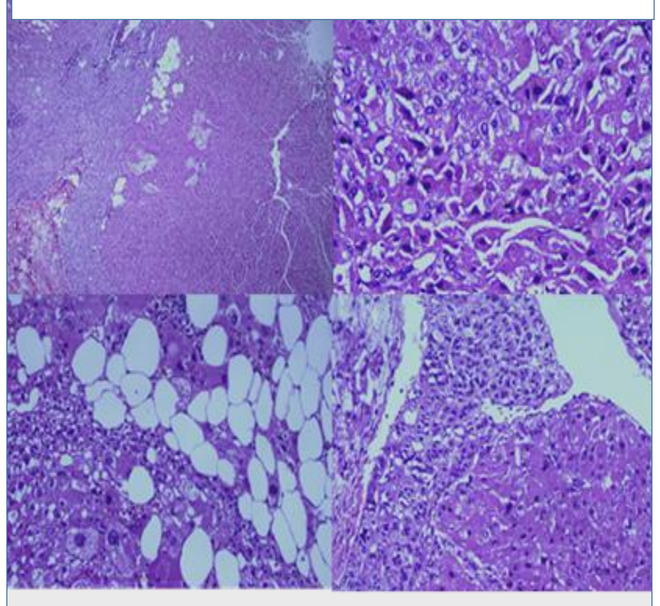

Figure 3. Microscopic features

DISCUSSION

PPNAD is a rare cause of CS, 50 percent is familial, associated with CNC. The disease usually presents before the age of 30 (4–44). The age of onset in our patient is 10.

Typical Cushing's syndrome is the frequent manifestation of PPNAD. The symptoms are usually mild that induce the late precise diagnosis, probably several years. Our patient was diagnosed 1.5 year after the onset.

PPNAD in children and adolescents can also manifest as variants of CS, like periodic CS or atypical CS that lead to difficulties in clinical diagnosis.

Total bilateral adrenalectomy was undergone for our patient . Actually, the treatment of choice for PPNAD is bilateral adrenalectomy to avoid the morbidity associated with CS. Unilateral or subtotal adrenalectomy is followed by recurrence.

The gross and histological appearance of PPNAD is characteristic and genetic studies are not necessary for a diagnosis.

Careful follow-up and screening, like annual echocardiogram and thyroid at

CONCLUSIONS

Primary pigmented nodular adrenocortical disease (PPNAD) is a rare cause of ACTH-independent Cushing's syndrome. Total bilateral adrenalectomy followed by hormone therapy is the optimal treatment. Periodic evaluation of CNC should be performed.

REFERENCES

1. Stewart PM: The adrenal cortex. Williams textbook of endocrinology Philadelphia: WB SaundersLarsen PR, Kronenberg HM, Melmed S, et al , 10 2003, 491-551.
2. Zografos et al. Bilateral adrenalectomy for Cushing's syndrome: Pros and cons. Journal of Medical Case Reports 2010, 4:230
3. Groussin L, Horvath A, Jullian E, et al. A PRKAR1A mutation associated with primary pigmented nodular adrenocortical disease in 12 kindreds. J Clin Endocrinol Metab 2006; 91:1943.
4. Powell AC, Stratakis CA, Patronas NJ, et al. Operative management of Cushing syndrome secondary to micronodular adrenal hyperplasia. Surgery 2008; 143:750.
5. Horvath A, Stratakis C. Primary pigmented nodular adrenocortical disease and Cushing's syndrome. Arq Bras Endocrinol Metab. 2007;51:1238–44.