# A CASE REPORT OF PRIMARY PIGMENTED NODULAR ADRENOCORTICAL DISEASE

## 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 familial, mostly associated with Carney complex (CNC). PPNAD may manifest by typical signs of CS, subclinical or cyclic CS. The optimal choice for treatment of PPNAD-induced CS is bilateral adrenalectomy followed by adrenal hormone 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; 3rd percentile for height).
- Laboratory tests: midnight salivary cortisol: 12.6 ug/dL (normal< 3.0 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- After surgery, hypertension improved, antihypertensive drugs were gradually withdrawn. Weight gain ceased. After 3 months: No hypertension, weight lost (3 kg/3 months), normal serum cortisol concentration.

## 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

5. Horvath A, Stratakis C: Primary pigmented nodular adrenocortical disease and Cushing’s syndrome. Arq Bras Endocrinol Metab. 2007;51:1236–44.