Is hyperaldosteronism a pathognomonic feature of conn's syndrome?

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Received: 15 November 2008 / Accepted: 16 November 2008
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Abstract A 10-year-old female child presented with resistant hypertension and hypokalaemia. There was no muscle weakness or periodic paralysis. The ratio of plasma aldosterone concentration to plasma renin activity was increased. MRI of the abdomen showed the right adrenal mass. The child underwent open right adrenalectomy. She had natriuresis in the postoperative period. She was normotensive without antihypertensive drugs at discharge and 2 months after surgery.

Keywords Conn's syndrome · Aldosterone · Renin · Hypokalaemia · Natriuresis

Introduction

Primary hyperaldosteronism with hypokalaemia accounts for about 0.7% of cases of hypertension in general population [1]. It may be due to unilateral adrenocortical adenoma (conn's syndrome 73%) or bilateral cortical hyperplasia (27%). Nearly 25 cases of childhood conn's syndrome have been reported in literature [2]. Here we report a case of conn's syndrome in a 10-year-old child.

Case report

A 10-year-old female child presented with resistant hypertension for 10 months duration (Fig. 1). Initially

Fig. 1 Patient's photograph, 10-year-old child

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Fig. 2 MRI abdomen showing the 3 cm mass in right adrenal

Fig. 3 Peroperative photograph showing capsulated right adrenal mass

the child had polyuria, polydipsia, headache, palpitation and there was no muscle cramps or weakness. Initial evaluation was done by her family physician. Diabetes mellitus was ruled out. Blood pressure was 170/100 mmHg. Blood urea and serum creatinine were normal. Serum electrolytes were sodium 141 mEq/l, potassium <1.5 mEq/l, chloride 93.2 mEq/l, bicarbonates 26.5 mEq/l. Antihypertensive drugs and potassium supplementation were started.

Further evaluation was done by nephrologist. USG abdomen and pelvis showed increased right renal echotexture. Dopplar showed no significant alteration in renal arterial flow.

CT scan of the abdomen showed 2.8 × 2.8 cm mass in right adrenal.

Serum cortisol was 8.5 µg%. (normal 5–25 µg%), Serum aldosterone was 418.8 pg/ml (normal for children 10–15 years is 40–480 pg/ml) with normal salt intake and measured after 2 hours of upright posture.

Plasma renin activity was 0.1 ng/ml/hr (normal supine 0.5–1.9 and standing 1.9–6 ng/ml/hr). Patient had symptomatic improvement and she was referred from the Institute of Child Health, Chennai to the Department of Endocrine Surgery, Madras Medical College for further management.

Patient was thin built, blood pressure was 130/80 mmHg and pulse rate was 84/min, patient did not have muscle weakness. There was no oedema. Mild hirsutism was present and external genitalia was normal. She was on tablet prazosin 1 mg bd, tablet alpramethylopa 250 mg tid, tablet apiranolactone 25 mg tid, tablet losartan 40 mg bd, tablet amlodipine 2.5 mg bd, syrup KCl 15 ml tid. Serum potassium was 3.3 mEq/l.

Plasma adrenocorticotrophic hormone (ACTH) was 41.6 pg/ml (normal 0–46), 24 hours urine metanephrine was 90 µg/day (normal <350).

Plasma aldosterone concentration in ng/dl to plasma rennin activity in ng/ml/hr ratio was 41.8:0.1 = 418. The ratio of plasma aldosterone concentration to plasma rennin activity of >30 is highly diagnostic of primary hyperaldosteronism [3].

MRI of the abdomen showed 3 cm mass in right adrenal and the left adrenal was normal (Fig. 2).

Patient underwent open right adrenalectomy (Figs. 3, 4). She had natriuresis in the early postoperative period. Urine

Fig. 4 Cut section of the specimen

Table 1 Showing the postoperative input and output chart

Postoperative day	Input ml	Output ml
1st	1,350	3,000
2nd	700	5,200
3rd	3,000	4,050
5th	3,400	3,900
7th	2,400	2,800
9th	1,900	2,200
11th	1,450	1,200

output exceeded the intake (Table 1). Blood pressure was maintained between 90/60 and 110/80 mmHg with normal saline infusion. This problem was settled in 10 days.

Patient was discharged on the 14th postoperative day. Blood pressure was 110/84 mmHg without antihypertensive drugs. Serum sodium was 132 mEq/dl and potassium was 3.3 mEq/dl. Adrenalectomy made this patient normotensive and normokalaemic. The long-term cure rate of hypertension by unilateral adrenalectomy for conn's syndrome averages 69% in reported series [4].

Histopathological examination revealed cortical adenoma (Fig. 5). Follow up after 2 months; blood pressure was 110/80 mmHg.

Fig. 5 Cortical adenoma, a low power view showing admixed smaller zona glomerulosa cells and clear cells resembling fasciculata cells

Discussion

Though conn's syndrome had been reported in a child of age 3½ years still it is a rare cause for hypertension in paediatric age group [5]. This patient had severe resistant hypertension with hypokalaemia (<1.5 mEq/l). Chronic hypokalaemia made the renal tubules resistance to vasopressin that resulted in polyuria [6]. Surprisingly there was no muscle weakness or periodic paralysis.

In paediatric age normal or increased plasma aldosterone concentration with suppressed plasma renin is highly diagnostic of primary hyperaldosteronism [6]. This patient had normal plasma aldosterone concentration and suppressed plasma renin. Plasma aldosterone concentration in ng/dl to plasma rennin activity in ng/ml/hr ratio was 41.8:0.1=418. The ratio of plasma aldosterone concentration to plasma rennin activity of >30 is highly diagnostic of primary hyperaldosteronism.

Normal plasma ACTH and normal cortisol levels ruled out the congenital adrenal hyperplasia (11-β-hydroxylase or 17-α-hydroxylase deficiencies). CT scan localized the tumour to right adrenal. The sensitivity of high resolution CT in locating adenoma ranges from 82 to 90% [7]. In this case contralateral adrenal pathology was not ruled out by CT. MRI scan of the abdomen revealed normal left adrenal gland.

Patient had natriuresis in the early postoperative period.

Conn's syndrome is diagnosed by clinical and biochemical investigations and not by histopathological examination.

Conclusion

High plasma aldosterone concentration is not always necessary to diagnose conn's syndrome in paediatric age group. Plasma aldosterone concentration to plasma renin activity ratio >30 is highly diagnostic of conn's syndrome.

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