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Severe hypertension in a ten-year-old boy secondary to an aldosterone-producing tumour identified by adrenal sonography

T Decsi, Gy Soltész, F Harangi, J Nemes,[1] M Szabó[2], A Pintér

Department of Paediatrics and First Department of Medicine[1], University Medical School, and Department of Radiology[2], County Hospital, Pécs, Hungary

Severe hypertension discovered incidentally in a 10 year-old boy was associated with persistent hypokalaemia and metabolic alkalosis. Primary hyperaldosteronism was diagnosed by demonstrating elevated plasma aldosterone levels and increased urinary aldosterone excretion with concomitant depressed plasma renin activity. Adrenal sonography identified a left adrenal adenoma which was removed surgically; normotension and normalization of plasma renin and aldosterone values ensued. This appeared to be the first use in children of sonography to identify adrenal adenoma and it is suggested to be the first step in the differential diagnosis of primary hyperaldosteronism

It is now well recognized that hypertension in childhood is not as rare as previously thought. The majority of children with mild or borderline hypertension have essential or primary hypertension, whereas children with severe symptomatic hypertension are likely to have underlying disorders including endocrine diseases with overproduction of hypertensive hormones.

We report on a child with severe hypertension discovered incidentally, who had a large aldosteronoma which was located by ultrasound and surgically removed.

Methods

Blood pressure was measured with the widest cuff that would fit between the axilla and antecubital fossa. For blood pressure measurements over the legs a large adult-size cuff (width, 16 cm) was used.

Plasma renin activities (PRA) were measured by Phadebas RIA-kit and plasma aldosterone levels by Aldok RIA-kit.

Report of Case

In a ten-year-old boy hypertension was recognized incidentally during physical examination before appendectomy. Blood pressures at admission were 170/130 mm Hg, 170/120 mm Hg, 150 mm Hg and 160 mm Hg measured over the right arm, left arm, rigth leg and left leg, respectively. No family history of hypertension or hypertension-relateddisease could be detected. Nothing abnormal could be found during routine physical and ophthalmoscopic examination. The chest X-ray was also normal, but the ECG showed signs of hypertrophy of the left ventricle.

Urinanalysis was normal, bacterial cultures of the urine were sterile.

TABLE I

Plasma renin activities and plasma aldosterone levels measured in supine and upright position before (1—3) and after (4) removal of the aldosterone-producing adrenal adenoma

No.	Sodium intake (mmol/day)	Potassium intake (mmol/day)	Plasma renin activity (ng/ml/h) in supine position	Plasma renin activity (ng/ml/h) in upright position	Plasma aldosterone level (pg/ml) in supine position	Plasma aldosterone level (pg/ml) in upright position
1.	130	50	0.02	0.022	218	291
2.	98*	80*	0.03	0.14	340	480
3.	153	80	0.02	0.045	375	360
4.	130	50	0.2	0.9	30	20

* Average of the preceding five days

Endogenous creatinine clearance was within physiological limits. Intravenous pyelography revealed normal kidneys and urinary tract.

Routine laboratory investigations showed persistent hypokalaemia (serum potassium 3.5—3.6 mmol/l) and metabolic alkalosis (pH 7.42—7.50; base excess 6.7—7.5 mmol/l) suggesting primary hyperaldosteronism as a cause of hypertension.

Plasma aldosterone level and plasma renin activity were measured at three levels of sodium intake in both upright and supine position (Table 1). On a standard hospital diet (sodium intake: 130 mmol/day), plasma aldosterone level was high (normal 50—175 pg/ml) and plasma renin activity was depressed (normal 0.3—2.0 ng/ml/h). After 5 days on low sodium intake (98.7 mmol/day) plasma aldosterone level increased further and plasma renin activity remained low. The non-suppressibility of the elevated plasma aldosterone was documented by measurement made on a high sodium intake (153 mmol/day) (Table I). The lack of physiological response to postural changes on normal and high sodium diet also suggested an "independent" aldosterone hypersecretion.

The ratio of daily aldosterone excretion to surface area is plotted against sodium excretion in Figure 1. Despite the high urinary sodium excretion (high sodium intake), aldosterone excretion remained high, suggesting inappropriate aldosterone hypersecretion.

On the basis of this information, the diagnosis of primary hyperaldosteronism (Conn-syndrome) was made; it was further corroborated by demonstrating the normalization of blood pressure after 3 weeks of spironolactone therapy (200 mg/day).

Normal plasma cortisol level, normal urinary cortisol excretion and normal VMA excretion indicated intact glucocorticoid and adrenomedullary functions.

Adrenal sonography (Picker International LS 3000) showed a round mass of approximately 4 cm in diameter over the left kidney. The mass could clearly be distinguished from both the kidney and the spleen (Fig. 2). No similar mass could be

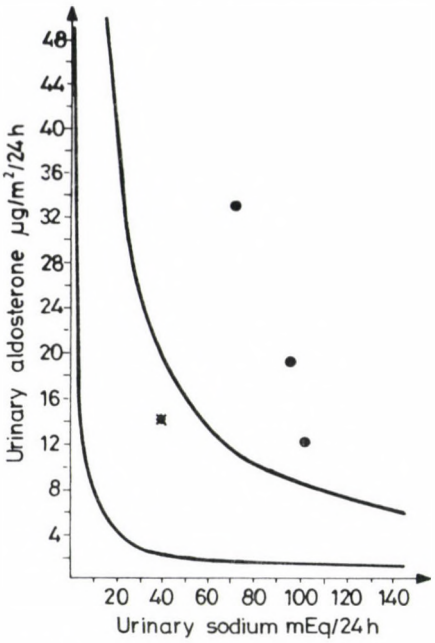

FIG. 1. Daily aldosterone excretion per surface area plotted against sodium excretion (5th and 90th percentile curves of normal subjects have been taken from New et al. [17]). The values of three subsequent preoperative determinations are shown by circles while the postoperative value by asterisk

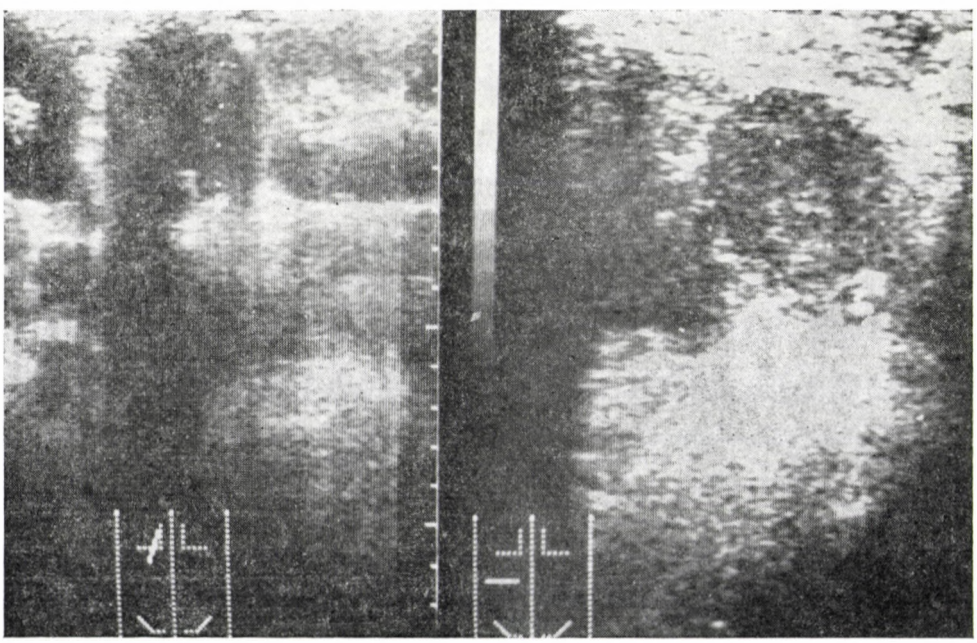

FIG. 2. Ultrasound scan of the region of the left suprarenal gland. A round mass of about 4 cm in diameter can clearly be distinguished from the kidney and the spleen

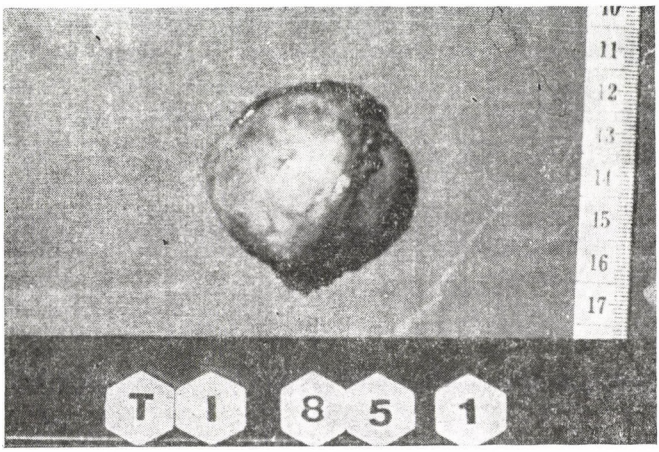

FIG. 3. Adenoma of removed left suprarenal gland. Weight: 27 g, diameter 4 cm

detected at the region of the right suprarenal gland.

At surgery a solid tumour weighing 27 g was removed from the left suprarenal gland (Fig. 3). Histological analysis of the tumour showed the characteristics of an adenoma. No sign of malignancy could be seen by histological examination.

The postoperative course was uneventful, neither the hypertension nor the hypokalaemia and metabolic alkalosis could be observed any more. PRA and aldosterone plasma levels (Table 1), as well as aldosterone excretion (Fig. 1) and also the physiological response of PRA to postural changes (Table 1) all returned to normal values.

DISCUSSION

Aldosterone overproduction is a rare but curable form of hypertension in paediatric age. It is suspected in a hypertensive patient who has persistent hypokalaemia and metabolic alkalosis. Hypokalaemia may remain asymptomatic as in our case, but it may also produce a variety of signs and symptoms which include fatigue, muscle weakness, paraesthesias, "periodic paralysis", polyuria, polydipsia, short stature, nocturia, etc. [8].

The diagnosis of primary hyperaldosteronism was readily established by the presence of elevated, nonsuppressible plasma and urine aldosterone and depressed plasma renin activity (low-renin hypertension).

The predominant adrenal pathology in childhood is bilateral adrenal hyperplasia [5, 6], aldosterone-producing tumour being exceedingly rare in children. To the best of our knowledge seven cases have been described up to now [4, 9]. As a subgroup of bilateral adrenal hyperplasia, a rare familiar form of hyperaldosteronism has also been described. The unique feature of this "dexamethasone suppressible hyperaldosteronism" is the com-

plete suppression of aldosterone secretion with dexamethasone administration.

There are several approaches to distinguish between adrenal hyperplasia and adenoma: photoscanning of the adrenal glands after ^{131}I-19-Iodocholesterol administration [1, 2, 16], determination of serum 18-hydroxycorticosterone level [10], computed tomography [11] and venography with determination of aldosterone level in samples obtained directly from the adrenal vein [7, 9]. Adrenal sonography has also been used to identify the adrenal lesion but has been considered difficult and often unsuccessful even in adults [11]. To the best of our knowledge there is no previous report of this method being used in children to identify adrenal adenoma. Furthermore, ultrasonic identification of the adrenal tumour made other complicated and not entirely harmless [12] diagnostic procedures superfluous.

On the other hand, it has to be emphasized that the tumour removed was unusually large even when compared with adrenal tumours in adults [1, 17]. It is probable that much smaller tumours could not always be detected by ultrasound technique. Nevertheless, since ultrasound evaluation is an entirely noninvasive method we suggest to use it as the first step in the differential diagnosis of primary hyperaldosteronism in childhood.

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G Soltész MD
József Attila u 7
7623 Pécs, Hungary