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Primary Hyperaldosteronism Due to Bilateral Adrenal Hyperplasia in a 13 Year-Old Boy

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Primary hyperaldosteronism is a syndrome which is characterised by hypertension, hypopotassemia and related symptoms.[1] Differential diagnosis of this syndrome from other causes of hypertension needs the determination of plasma renin activity and 24 hours urinary aldosteron excretion.[1,2]

In pediatric age group the etiology of primary hyperaldosteronism is generally bilateral adrenal hyperplasia, In the literature the number of reported cases so far is ten.[3] Adrenal adenoma and carcinoma are very few in number.[4,5]

The histopathology of adrenal shows hyperplasia of the zona glomeru losa, fasciculata or both.

The treatment of bilateral adrenal hyperplasia consists of bilateral adrenalectomy. Also spiranolacton can be used for hypertension. In some form of this syndrome the benefit of dexamethasone is reported. The purpose of this report is to describe a case of juvenile primary hyperaldosteronism due to nodular hyperplasia, and to present the diagnostic and therapeutic measures.

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Case Report

A. Ö., a 13 year-old boy was first admitted to Hacettepe University Medical Center on January 1.1977 for the evaluation of polyuria, Polydypsia, head ache and episodic weaknees of his extremities. These symptoms were present for 6-7 years. His past history revealed that 6 years ago he began driking about 2-3 liters of water a day and also to urinate 12-13 times a day without any irritative or obstructive symptoms.

5 years ago he began complaining of the weaknesss of his arms, hands and legs, which were episodic and seen in the morning. At the time of admission he was on no medication The physical examination revealed a blood pressure of 135/105 mm Hg in both arms with no postural change. Pulse rate was 75/min, weighth 27 kg, Height 135 cm. Fundus was normal; there were no detectable murmurs.

Laboratory studies revealed: Urinary spesific gravity of 1003, sterile urine culture, PH 6 no sugar, no protein microscopic examination revealed 2-3 epitheliall cells. BUN: 29 mg/dl, Creatinin: 1.4 mg/dl, Na: 144 mEq/lt, K: 1.8 mEq/lt CL: 111 mEq/lt, Co2: 29 mEq/lt, P: 2.8 mg/dl, Ca: 10 mg/dl, PCO_2: 32 mmHg blood PH: 7.44, Total protein: 6.4 gr/dl, Albumin: 4.4 gr/dl.

IVP was nokmal, X-Ray of the chest revealed cardiomegaly and left ventricular prominence. Skull X-ray was found normal Retroperitoneal air insufflication was found normal.

24 hours urine output was 6 liters. During water deprivation for 8 hours the specific gravity of the urine increased to 1009. Pitressin had no effect on 24 hours urine volume and concentration. After a low sodium diet (Na: 10 mEq/day) for three days, blood was withdrawn in supine position for plasma renin activity and was found 0.15 ng/ml/hr. After a high sodium diet (Na: 150 mEq/day, K: 80 mEq/day) for three days, 24 hours urine was collected for aldosteron determination and was found as 8.4 micro gr/24 hr. During the high sodium regimen his blood pressure increased up to 190/150 mmHg.

A spiranolactone therapy (100 mg/day) was recomended Two months later he had no complaint, electrolytes were found normal and urine spesific gravity was 10.13. His blood pressure was found 160/120. In June 1977 the patient underwent bilateral adrenal exploration. At surgery the righth adrenal gland was found to be enlarged and nodular, the left one was normal Right adrenalectomy and renal biopsy were performed. Microscobic sections revealed noduler hyperplasia especially

In zona fasciculata. The renal biopsy revealed medial hypertorpy of the medium sizedrenal arteries and mild hyalinization in a few glomeruli. Also in proksimal tubules there was vacualisation secondary to hypokalemia.

In the post operative period the blood pressure still remained at 160/120 mmHg, so we recomended spiranolactone 100 mg/day. The patient is entirely asympthomatic and is well maintained on spiranolactone 100 mg/day.

Discussion

The combination of hypertension, hypokalemia, pitressin resistant polyuria, suppressed plasma renin activity and increased urinary aldosteron excretion estabilshed the clinical diagnosis of primary hyperaldosteronism in our case.

The pathologic study also confirmed the clinical diagnose. In the literature we could only found ten cases of primary hyperaldosteronism due to adrenal hyperplasia in childhood.[3] Their age were between 9-23 years old. The first case of van Buchen was (a) 17 years old boy with the hyperplasia of both zona glomerulosa and retucularis. After this case 9 additional cases were pulbished. Four of them showed normal histology, 2 showed hyperplasia of Z. G and Z. F. one showed hiperplasia of Z. G. and three showed noduler hyperplasia. Five of them had bilateral adrenalectomy, and had subtotal adrenalectomy. All the results were good. The renal biopsies revealed arteriolar sclerosis and hypokalemic nephropathy in two cases.

Our case exhibited all the features deseribed in previous cases. Only the surgical therapy was different. Because of the normal appearence of the left adrenal we did not perform bilateral adrenalectomy.

With the addition of spiranolactone 100 mg/day, his bloodpressure is under control now and also he doesnt need glucocorticoid and mineralecorticoids.

Although the current therapy is bilateral adrenalectomy we are able to control hypertensionwith partial adrenalectomy plus spiraoloactone, and also we can not be sure that whether the hypertension would be controlled after bilateral adrenalecotmy because of the renovascular changes. Inorder to decide for the removal of the left adrenal it seems to be better to wait and control blood pressures frequently.

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