A CASE OF PHEOCHROMOCYTOMA DIAGNOSED AS ADRENAL INCIDENTALOMA

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BACKGROUND

Adrenal masses larger than 1 cm determined incidentally during radiological examinations or abdominal laparotomy are called adrenal incidentalomas. Incidentalomas were shown to be seen more than expected according to the serial autopsy findings. In various series, its prevalence was reported to be 1-8.7%. Incidentaloma prevalence increases with age; in childhood and adolescence it is nearly 0.2%, whereas in the advanced ages it increases up to 10%.

There are two main problems that should be solved when encountering adrenal incidentaloma. The first of these is to decide whether the adrenal mass is benign or malignant and the second is to determine whether the mass is hormonally active or not.

OBJECTIVES

By means of this rare case, we aimed to focus on the approach to adrenal masses in childhood in general and on the approach to pheochromocytoma in specific.

METHOD

A case of pheochromocytoma, which was diagnosed as incidentaloma in childhood, is presented.

RESULTS

A 17-year-old male patient was admitted to the hospital with complaint of progressive weight gain. Because of abnormal liver function tests (AST: 118 U/L ALT: 115 U/L), abdominal USG was applied, and in addition to grade 2 steatohepatitis and hepatomegaly, it showed hypoechoic mass with regular boundaries and in dimensions of 60x51 mm, localized in the left adrenal gland. The abdominal CT revealed a mass that consisted of areas of cystic necrosis and had dense heterogeneous contrast-enhancement, localized in the left adrenal gland. The patient was diagnosed with adrenal incidentaloma. The dimensions of the tumor and radiological visualization help to determine if the mass is benign or malignant. The mass detected was larger than 6 cm, its appearance was heterogeneous, and it involved calcification and cystic necrotic areas. In the CT without contrast, density was 20 HU; due to hypervascularization its contrast content was dense, contrast did not disappear rapidly after being applied, and washout was <60% after 10 minutes. With these findings, the mass was not in accordance with adenoma.

RESULTS

The patient was reevaluated clinically, and clinical signs due to excess cortisol secretion such as moon face, plethoric face, purple striae, easy bruising or hyperglycemia were not detected. Hypokalemia due to excess aldosterone secretion, polyuria, or polydipsia did not exist. Findings due to excess catecholamine secretion such as episodic or continuous hypertension, palpitation, headache, sweating episodes, pale or flushing face, anxiety, tremors also did not exist. The case was accepted to be clinically asymptomatic. We evaluated the adrenal tumor for its hormonal activity, and the laboratory analysis results are shown in Table 1.

Blood pressure levels of the case were within normal ranges during the controls. Since hypertension can be associated with most of the adrenal tumor cases (pheochromocytoma, Cushing syndrome, and hyperaldosteronism), his blood pressure was monitorized for 24 hours. Selective alpha-1 antagonist doxazocin therapy was prescribed due to determination of hypertensive values. After preparing the patient preoperatively, left surrenalectomy was performed. The pathological analysis was in accordance with pheochromocytoma.

The patient was screened in terms of accompanying syndromes (MEN type 2, Von Hippel-Lindau Disease, Neurofibromatosis type 1, familial pheochromocytoma /paraganglioma syndromes) and no positive findings were detected.

Table 1. Laboratory analysis to evaluate the etiology of tumor

Prediagnosis	Laboratory Tests Performed
Subclinical Cushing syndrome	Plazma ACTH level: 29.1 pg/ml (N: 10-60) Serum cortisol level: 13.03 µg/dl (N: 4.3-22.4) Night time cortisol: 3 µg/dl (N: 3.0-16.7) 24 hour urine free cortisol:40 µg/gün (N:11-84)
Primary hyperaldosteronism	Serum K: 4.1. mEq/L (N: 3.5-5) Renin: 15.7 pg/ml (N: 5-33) Aldosteron: 80 pg/ml (N: 30-313)
Androgen secreting tumor	DHEA-S: 209 µg/dl (N: 108-441) Testosteron: 313 ng/dl (N: 300-1200) 17-OH-Progesteron: 0.8 ng/ml (N: 0-2)
Pheochromocytoma	Catecholamine and its metabolites in 24 hour urine 3-Methoxytyramine: 170.95 µg/day (83-459.8) Metanephrine: 5.39 µg/day (42-347.7) Epinephrine: 18.96 µg/day (4-20) Vanilmandelic acid (VMA): 39.78 µg/day (1.6-7.3) 5-hydroxy indole acetic acid: 15.8 µg/day (0.5-8.2) Norepinephrine: 7598.39 µg/day (23-105) Dopamine: 1491.64 µg/day (190-450) Normetanephrine: 4543.75 µg/day (72-659) Homovanilic acid (HVA): 10.8 µg/day (1.4-8.8)

CONCLUSION

The present case is a rare case of pheochromocytoma that was diagnosed as adrenal incidentaloma in childhood. Pheochromocytoma and Cushing's syndrome should be excluded in all cases with adrenal incidentaloma. Primary aldosteronism should be excluded in patients with hypertensive and/or hypokalemic episodes.

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