CASE REPORT

A Child with Pheochromocytoma and Malignant Hypertension.

by

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Abstract

Herein we report a boy of 13 years old who suffered from pheochromocytoma with malignant hypertension. No metastasis was found and surgical approach was done successfully. Histopathologic examination showed pheochromocytoma of the left suprarenal gland with pleomorphism in the primary tumor.

Postoperatively the blood pressure returned to normal within one month and remained so within 3 years follow up.

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Introduction

Pheochromocytoma was first described as a tumor in 1886 by Frankel. However, it was not until 1912 when the nomenclature pheochromocytoma was established by Pick. The first correct clinical diagnosis of paroxysmal hypertension was diagnosed by Vaquez and Donzelot in 1966.

Pheochromocytoma is a rare tumor with an incidence described by Hume (1966) of 1 in each 1000 autopsies. Malignancy was reported by Remine et al., (1974) 13,1% and Petrovsky & Krylov (1970) 8 — 11%. The tumor was inherited by an autosomal dominant gene. Malignancy of pheochromocytoma depends on the clinical course characterized by recurrence or metastasis and pathologic demonstration of the tumor in area where chromaffin tissue does not normally exist, such as lymph nodes.

Case Report

M. 13 years old, an Indonesian boy was admitted to the Subdivision of Nephrology, Department of Child Health, Dr. Cipto Mangunkusumo Hospital, Jakarta on March 5, 1978 (Fig 1). The chief complaint on admission was severe headache, sweating, palpitation and difficulty in sleeping for 4 days. The family history showed that the father and one of the brothers of this child had the same complaints and died suddenly when they were asleep at night. During hospitalization the blood pressure ranged between 120/100 mmHg and 240/190 mmHg, despite antihypertensive drugs. The excretory urogram (IVP) showed no abnormality and renal angiography revealed left suprarenal hypervascularization and left renal artery stenosis (Fig 2).

The renogram showed disturbances of the left renal function.

The regitine test was positive. The VMA of the urine was 29,4 mg/24 hours and fasting blood sugar 164 mg/dl Funduscopic examination showed no retinopathy and there was also no enlargement of the heart an the chest X-ray picture.

During hospitalization he got priapism the cause of which was difficult to explain and needed surgical correction. During surgical intervention under general anesthesia, hypertensive crisis appeared but could be controlled by regitine i.v.

Exploration of the tumor was done through a left flank incision. A large left suprarenal tumor was found and left adrenalectomy was performed. Five days before exploration he received alpha-blocker (prazosin) and beta-blocker (propanolol). During operation the blood pressure increased to over 240/190 mmHg but could be controlled by regitine i.v. and no hypotension occurred after surgery.

The measurement of the tumor was $5 \times 3 \times 3$ cm and weighed 50 gm. (fig. 3). Histopathologic examination showed large tumor cells with basophilic cytoplasm, granular and nuclear pleomorphism in every portion. A part of the

tumor cells showed positive chromaffin reaction. At the border of the tumor a thin adrenal cortex was seen. In conclusion phaeochromocytoma of the left adrenal gland was confirmed (fig.4).

The previous signs and symptoms of the child disappeared after operation and the blood pressure decreased until normal within one month and remained so until 3 years of follow up.

Discussion

The diagnosis of pheochromocytoma is usually made clinically. The most common symptoms are headache, sweating, nausea and vomiting, visual changes, weight loss, weakness, abdominal pain, nervousness, palpitation, fatigue and pallor. There is also a high incidence of polydipsia and polyuria. Preoperatively our patient showed headache, sweating, nausea, weight loss, weakness, pallor and polyuria suggesting pheochromocytoma as the cause of hypertension. The fasting blood sugar was 164 mg/dl. Hume (1960) reported that 40% of his patients showed fasting blood sugar elevation to over 120 mg/dl. The IVP of our case revealed no abnormality. IVP examination can demonstrate the location of the tumor by showing distortion of the renal pelvic or displacement of the kidney. The location of the tumor was found by renal angiography which showed hypervascularization of the left suprarenal area. Another method was described by Mahoney et al., (1967) by vena caval catetherization with plasma catecholamines determination on blood samples drawn at intervals as the catheter is passed along the vena cava.

In our case the left renal artery was stenotic. Preoperatively, it was questionable whether it had any contribution to the elevation of the blood pressure. But since the blood pressure returned to normal postoperatively, it was assumed that it had no significant meaning to the hypertension in this case. Renogram revealed disturbances of the left kidney function which could be due to the stenosis of renal artery. Pheochromocytoma with reversiblo renal artery stenosis was described by Volick et al., (1978). Stenosis were frequently attributed to compression or stretching of the renal artery by the tumor. The stenosis regressed following treatment with propanolol and phenoxybenzamine suggesting an adrenergic mechanism as the cause.

Monitoring of the blood presure revealed a range between 120/100 mmHg and 240/190 mmHg. VMA of the urine revealed 29,2 mg/24 hours and regitine test was positive. In this case the hypertension was assumed to occur acutely since there was no ophthalmic retinopathy and enlargement of the heart.

Successful operation of pheochromocytoma is greatly enhanced by intensive preoperative and intraoperative care. Intraoperative manipulation of the tumor will often result in release of large amount of catecholamines and the necessary precautions should be taken, using alpha and beta adrenergic blocking agents (Daughtry et al., 1977). For the

same reason this patient received preoperatively an alpha (prazosin) and beta blocker (propanolol) five days previously. During operation blood pressure was elevated to over 240/190 mmHg but could

Removal of the tumor is often followed by hypotensive crisis due to the elimination of the chronic catecholamines stimulation of the peripheral arterioles in volume depleted patients. Fortunately it did not happen to our patient. Preoperative transfusion was given to prevent the possible hypotensive crisis as suggested by DeOreo et al., (1974).

Postoperatively the blood pressure return to normal within one month. If persistent hypertension occurred it could be due to the left renal artery stenosis or the possibility of another ectopic tumor located elsewhere.

As in our case about 90% of the tumors were situated in the adrenal gland. Remine et al. (1974) reported 124 intraadrenal tumors, 70 (56,5%) were in the right gland and 48 (38,7%) in the left, 6 (4,8%) bilaterally.

Adrenal tumors vary in size and weight. The measurement of the tumor in this case was 5 × 3 × 3 cm and weighed 50 gm. Sherwin (1959) reviewing 96 patients with adrenal pheochromocytoma reported that the average weight of these tumors was 90 gm. Gifford et al. (1964) reported the weights of the tumors ranged between 5 to 500 gm while Daughtry et al., (1977) reported also a giant pheochromocytoma weighing 1.150 gm and measuring 17 × 15 × 8 m.

Remine et al. (1974) reported that the recurrence rate of the tumor after surgical treatment was 9,8% and five years survival subsequent to treatment for the benign tumor was 96% and for malignant tumor 44%. Recurrence developed with a median distribution of 5.6 years after operation (Remine et al., 1974) while Mahoney and Harrison (1977) reported an interval of 8 years and recommended that all patients with pheochromocytoma should have a detailed follow up every 6 months for 15 years. In our case the blood pressure remained normal until 3 years follow up.

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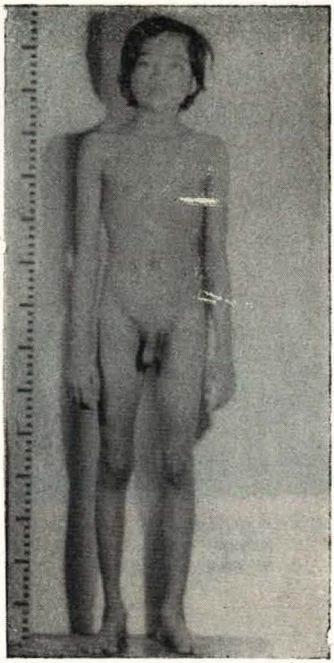

FIG 1. *Patient: M. an Indonesian boy, 13 years old before operation.*

FIG 2. *Renal angiography showed left suprarenal hypervascularization and left renal artery stenosis*

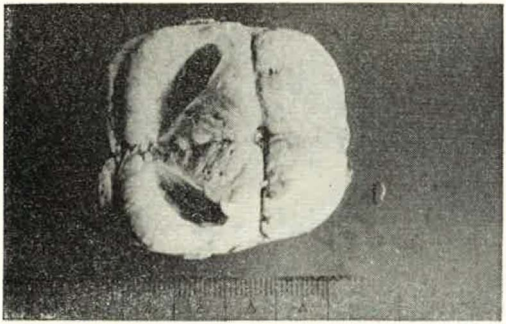

FIG. 3. *Left adrenal tumor, the measurement was* 5 × 3 × 3 *cm and weighed* 50 *gm. (split).*

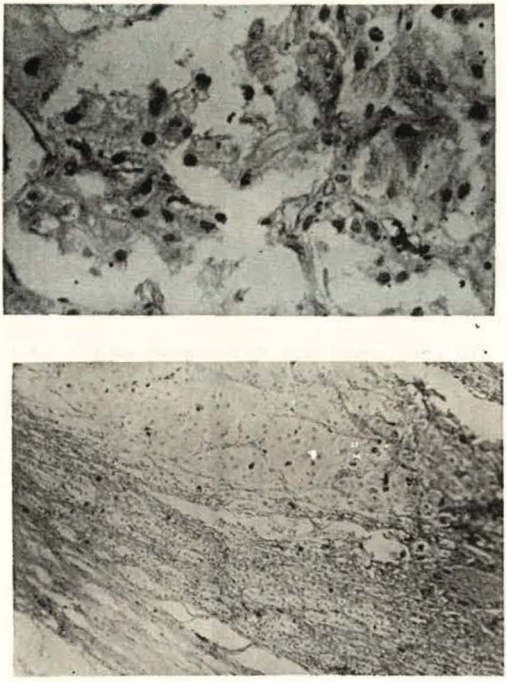

FIG. 4. *Histopathologic examination showed large tumor cells with basophilic cytoplasm, granular and nuclear pleomorphism in every portion. A part of the tumor cells showed positive chromaffin reaction and at the border of the tumor a thin adrenal cortex was seen.*