A CASE OF NORMOTENSIVE PHEOCHROMOCYTOMA MASQUERADING AS A DILATED CARDIOMYOPATHY

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A case of right adrenal pheochromocytoma masquerading as a dilated cardiomyopathy is described. This patient was normotensive throughout 8 years of observation. Hyperglycemia and an abnormal glucose tolerance test were a clue to the diagnosis which was confirmed by the findings of increased plasma and urinary epinephrine and norepinephrine values. In patients with dilated cardiomyopathy the possibility of a pheochromocytoma should be considered.

INTERMITTENT or sustained hypertension is a common symptom of pheochromocytoma. Although associated myocardial lesions known as catecholamine myocarditis or cardiomyopathy1-3 were reported to be found in more than half of the patients3 a cardiac state simulating dilated cardiomyopathy with congestive heart failure is very rare4-11 We report a patient with pheochromocytoma but without hypertension presenting as a dilated cardiomyopathy.

CASE REPORT

A 57-year-old man initially visited our hospital 8 years ago (1977) with the chief complaint of palpitations when his blood pressure was 125/90 mmHg and heart rate 65/min. Neither an electrocardiogram (ECG) nor chest X-ray (CTR = 0.38) disclosed any abnormalities (Figs. 1, 2). Two years later (1979) he was admitted to another hospital for the evaluation of syncope and was given a tentative diagnosis of cardiomyopathy. In 1982 he readmitted to a third hospital for the evaluation of palpitations. At that time ECG showed inverted T waves in leads I, II, III, aVL, and V5,6, with frequent ventricular premature contractions. A chest X-ray revealed a cardiothoracic ratio of 0.57. The findings of a cardiac biopsy was reported to be compatible with dilated cardiomyopathy.

In 1984 he was readmitted to our hospital for further evaluation and therapy of his congestive heart failure and frequent ventricular premature

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Fig.1. A chest X-ray in June, 1977 showing no abnormalities.
Fig. 2. An electrocardiogram in June, 1977 is within normal limits.

Fig. 4. An electrocardiogram showing sinus tachycardia, left axis deviation, intraventricular conduction disturbances, poor R/S progression and frequent ventricular premature contractions.

Fig. 3. A chest X-ray in February, 1985 with moderate cardiomegaly, pulmonary congestion, interstitial edema and right sided pleural fluid.

Contractions. On physical examination the patient appeared chronically ill, the heart rate was 90 and blood pressure 100/50 mmHg. No lymphadenopathy was found. No jugular venous distention was noted. The heart was moderately enlarged and a Grade 3 holosystolic murmur was heard at the cardiac apex. Fine crackles were present posteriorly. Abdominal examination was negative and there was no peripheral edema. A chest X-ray showed moderate cardiomegaly (CTR = 0.64), prominent pulmonary vasculature, interstitial edema and pleural effusion on the right (Fig. 3).

The ECG showed a sinus rhythm with a heart rate of 105, low voltage in limb leads, left atrial overload, left axis deviation, intraventricular conduction defects, poor R/S progression and frequent ventricular premature contractions (Fig. 4). An M-mode echocardiogram disclosed prominent left atrial enlargement (LAD = 52 mm), right ventricular enlargement (RVD = 45 mm), left ventricular enlargement (LVDd = 677 mm, LVDs = 588 mm), hypokinesia of the intraventricular septum (wall thickness = 10 mm) and left ventricular posterior wall (thickness = 10 mm). There was a posterior wall excursion of 5.7 mm, a left ventricular ejection fraction of 0.35 (Gibson's formula), a mean Vcf of 0.28 circ/sec, and decreased EF slope of anterior mitral leaflet (30 mm/sec). All the findings on echo suggested a dilated cardiomyopathy (Fig. 5).

Since a fasting blood sugar was 180 mg/dl, an oral glucose tolerance test (75 g) was done which disclosed a fasting blood glucose of 150 mg/dl, 373 mg/dl at 1 hr, 213 mg/dl at 2 hrs, 34 mg/dl at 3 hrs with IRI 757 μU/ml at 1 hr and IR1 1000 μU/ml at 2 hrs. An abdominal echogram and computed tomography revealed a tumor of the right adrenal gland with a maximal diameter of 64 x 67 mm (Fig. 6). The epinephrine level was significantly increased to 3180 pg/ml in plasma, 755 μg/day in urine. The norepinephrine level was 5150 pg/dl and 1230 μg/day, respectively. The urinary vanillylmandelic acid increased to 51.0 mg/day. The serum dopamine level, however, stayed within normal limits (< 200 pg/ml).

Although the diagnosis of pheochromocytoma was apparent, the patient had not been troubled by hypertension either paroxysmal or persistent, or by symptoms of headache, sweating or nervousness. His systolic blood pressure had ranged from 90 to 140 mmHg and diastolic from 40 to 90 mmHg. His heart rate ranged from 65 to 105
Fig. 5. An M-mode echocardiogram showing an enlargement of all four chambers, hypokinesis of the ventricular septum and left ventricular posterior wall and decreased E-F slope of the anterior mitral leaflet.

Fig. 6. a: Abdominal computed tomography showed a mass anterior to the right adrenal gland. b: Abdominal echogram showing a mass derived from the right adrenal gland.

in more than 155 readings throughout 8 years. He was discharged on the 17th hospital day on conventional congestive heart failure therapy in addition to phenoxybenzamine. Although surgical excision of the pheochromocytoma had been considered, it had not been done because patient's cardiac condition was thought to be too poor to be able to tolerate major surgery. After
Normotensive Pheochromocytoma with DCM

discharge, his cardiac state worsened and he died of congestive heart failure 2 months later.

Needle necropsy of the right adrenal tumor showed cells composed of eosinophilic and granular cytoplasm and nuclei of varying sizes. The argyrophilic nature of these cells was detected by Grimelius staining. Although the necropsy specimen of the heart was not sufficient, microscopic examination revealed enlarged muscle cells and mild fibrosis.

DISCUSSION

Absence of Hypertension

Pheochromocytoma secretes norepinephrine and epinephrine with combined alpha- and beta-adrenergic stimulation which usually results in labile or sustained systemic hypertension. By contrast, only a small number of patients have been reported to be normotensive. The absence of hypertension in some patients with pheochromocytoma has been explained as follows: (1) predominant secretion of epinephrine, (2) concomitant dihydroxyphenylalanine (dopa) secretion, (3) inactivation of the norepinephrine within the tumor, (4) myocardial failure, and (5) tolerance of tissue receptors to circulating catecholamines.

In the present case, norepinephrine secretion was predominant as shown by plasma and urine values and the plasma dopamine value was within normal limits. The catecholamine excretion for norepinephrine was high enough to rule out inactivation within the tumor. Since this patient had continued to be normotensive throughout the 8 years of observation, myocardial failure itself seems not to have played a role at the beginning of the disease. Therefore, in light of the above observations, tolerance of tissue receptors to circulating catecholamines may have occurred in this patient. Although this tolerance may be present in some patients with pheochromocytoma, the exact mechanism of maintaining normal blood pressure in some norepinephrine-secreting pheochromocytomas is still controversial.

Catecholamine Cardiomyopathy

In experimental animals, repeated injections of catecholamines can induce left ventricular dilatation and hypertrophy which is known as catecholamine myocarditis or cardiomyopathy. In man, excessive secretion of catecholamines, as shown in pheochromocytoma, has been said to result in equivalent myocardial lesions. This may induce ST-T changes, serious arrhythmias, congestive heart failure, shock, myocardial infarction and significant cardiac hypertrophy or enlargement mimicking cardiomyopathy. There have been a few reported cases presenting as hypertrophic cardiomyopathy, with or without outflow tract obstruction, which was ascribed to the hypercontractile state induced by catecholamines.

On the other hand, we found 4 reports of patients with pheochromocytoma who were suspected of having a dilated cardiomyopathy although none of them had echocardiograms. Garcia and Jennings reported a case of pheochromocytoma with congestive heart failure and severe postcapillary pulmonary hypertension, which showed remarkable clinical and hemodynamic recovery after removal of the tumor. Interestingly enough, three of these patients were relatively normotensive. In three of them, disappearace of the symptoms of congestive heart failure and a prominent decrease of the cardiac silhouette on chest X-ray were noted after resection of the tumor, supporting the view that the myocardial lesions may be due to the direct toxic effect of catecholamines on the heart.

REFERENCES


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