Normotensive cardiomyopathy and malignant hypertension in phaeochromocytoma

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Summary

A patient with two different presentations of phaeochromocytoma is described. She initially presented with normal blood pressure and heart failure following a prolonged feverish prodrome. A provisional diagnosis of myocarditis or early congestive cardiomyopathy was made and she improved with digoxin and diuretics. Eighteen months later, after a period of normotension free from heart failure, she developed malignant hypertension with recurrence of heart failure. A phaeochromocytoma was surgically removed, with return to normal of blood pressure and cardiac status. It would seem that the initial presentation of the phaeochromocytoma was a catecholamine-induced myocarditis without hypertension and this resolved with the subsequent development of malignant hypertension. The possible mechanisms responsible for this are discussed and it is concluded that phaeochromocytoma should be considered in patients who have heart failure and persistent features of myocarditis.

Introduction

Phaeochromocytomas are rare tumours with a constellation of clinical features dominated by hypertension, due to the constant or paroxysmal release of pressor catecholamines (Engleman, 1977); myocarditis and congestive heart failure may also directly result from the action of these agents (Vliet, Burchell and Titus, 1966). The case presented illustrates that a normotensive patient with phaeochromocytoma and congestive heart failure may produce diagnostic problems.

Case report

A 30-year-old married woman who was born in India presented in September 1978 with a 6-week history of dry cough, profuse nocturnal sweating, exertional dyspnoea and ankle oedema. There was

no family or past history of note. She had a swinging pyrexia (37-39.6°C), persistent tachycardia (heart rate varying from 106 to 138 beats/min) and blood pressure between 105/65 and 122/78 mmHg on more than twenty occasions. Jugular venous pressure was raised 7 cm, and third and fourth heart sounds were heard but no other abnormalities were noted on physical examination. Chest radiography showed an enlarged cardiac silhouette and the electrocardiogram showed inferior and lateral lead ST segment depression and T wave inversion. An echocardiogram showed normal wall and septal thickness, enlargement of the left atrium and ventricle with reduced shortening. The activity of aspartate aminotransferase was raised to 245 i.u./l (normal 45 i.u./l), metanephrine excretion was not estimated and all other biochemical, endocrinological and microbiological investigations were normal. A provisional diagnosis of viral myocarditis or early congestive cardiomyopathy was made, and she was treated with digoxin and diuretics with gradual symptomatic improvement and no change in blood pressure. No rise in viral antibody titres was shown.

She was regularly reviewed and became asymptomatic by November 1978 with a normal chest radiograph. Her blood pressure was recorded until February 1979 at between 112/74 and 128/82 mmHg. However, in April 1979 heart failure recurred and she was noted to be very anxious with a blood pressure of 205/142 mmHg, and to have retinal exudates, haemorrhages and papilloedema. Investigation revealed raised urinary metanephrine excretion at 42.4 µmol/l 24 hr (normal 9.3 µmol/24 hr) with metadrenaline at 980 µg/g of creatinine (normal 130), and nor-metadrenaline at 2964 µg/g of creatinine (normal 220). Arteriography showed a left suprarenal mass. A 70 g phaeochromocytoma was excised and subsequently the blood pressure,

radiographic heart size, electrocardiogram and echocardiogram returned to normal and she has remained well since.

Discussion

Hypertension with or without retinal changes, myocarditis or congestive heart failure are well recognized features of phaeochromocytoma (Engleman, 1977). An interesting aspect of this case was the development and resolution of heart failure with entirely normal blood pressure before the development of malignant hypertension. The diagnosis of viral myocarditis or early congestive cardiomyopathy is often difficult to make but the clinical features of pyrexia, tachycardia and heart failure following a feverish prodrome are highly suggestive even in the absence of rising viral titres (Braunwald, 1980); However, in the light of subsequent events, this initial episode may have been due to a catecholamine-induced myocarditis.

Excess catecholamine secretion may induce hypertrophy, disseminated focal degeneration and necrosis of myocardium (Vliet *et al.*, 1966). This may result in acute and chronic heart failure, occasionally in the absence of hypertension (Baker *et al.*, 1972). In the case presented, electro- and echocardiograms revealed no hypertrophy and this illustrates that, although left ventricular hypertrophy is described as a feature of catecholamine myocarditis (Vliet *et al.*, 1966), this is due to associated hypertension. It has been recently shown that myocardial hypertrophy is absent in some cases of malignant hypertension due to other causes (Shapiro, Mackinnon and Beevers, 1981), and this patient confirms that the malignant phase may be of rapid onset and not

preceded by a prolonged non-malignant phase. Excessive catecholamine action is almost certainly the cause of left ventricular disease in phaeochromocytoma and is one hypothesis for the evolution of congestive cardiomyopathy (Swedberg *et al.*, 1980).

Thus, this case has unusual features with two different presentations of phaeochromocytoma possibly owing to variations in the amount and relative concentrations of adrenaline and noradrenaline, and integrates some aspects of the development and resolution of congestive cardiomyopathy and non-hypertrophic malignant hypertension. There were no specific features to indicate the aetiology of the initial heart failure. However, the symptoms and signs of active myocarditis were rather prolonged and it is concluded that the diagnosis of phaeochromocytoma should be considered in cases of heart failure of unknown aetiology.

References

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