Reno-vascular disease in polycythaemia rubra vera

BEVERLEY BIGGS
M.B.B.S.

J. L. ANDERTON
M.B., F.R.C.P.E.

Western General Hospital, Edinburgh

Summary

Hypertension and polycythaemia rubra vera commonly occur in the same patient. This is regarded as coincidence and the hypertension does not respond to correction of polycythaemia. We report a case of renal artery stenosis occurring simultaneously with polycythaemia rubra vera and suggest that renovascular occlusive disease should be excluded in such patients with refractory hypertension.

KEY WORDS: hypertension, renal artery stenosis, polycythaemia rubra vera.

Introduction

Erythrocytosis is now a well-known association of renal artery stenosis. In 1962, Penington postulated that factors which impaired renal perfusion should lead to increased erythropoietin secretion and secondary erythrocytosis. The first report in 1965 (Luke *et al.*) and subsequent papers (Hudgson, Pearce and Yates, 1967) have suggested a similar pattern of erythrocytosis and hypertension secondary to increased erythropoietin and renin secretion in response to renal ischaemia. Each of these patients has been shown to have secondary polycythaemia. We present an unusual case of renal artery stenosis occurring in a patient with polycythaemia rubra vera.

Case report

A 47-year-old man was admitted to hospital in November, 1981 for control of hypertension. He was first found to be hypertensive in 1979. Subsequent treatment with propranolol and hydralazine on 3 occasions resulted in gangrenous toes. The patient had never smoked and had no other history of note. There was no family history of hypertension.

On clinical examination, he appeared plethoric and was hypertensive (blood pressure 180/115 mmHg). A loud epigastric bruit was audible. Palpation of the abdomen revealed 10 cm of splenomegaly. The left fifth toe was gangrenous.

Initial investigations showed haemoglobin (Hb)

18.3 g/dl; red cell count 6.89×10^{12} /litre; white cell count (WCC) 11.7×10^9 /litre; platelets 505×10^9 /litre; erythrocyte sedimentation rate 1 mm in 1 hr; plasma electrolytes normal; urea 8.0 mmol/litre, and creatinine 0.16 mmol/litre. Urinalysis was negative and the creatinine clearance was 54 ml/min.

True polycythaemia was confirmed by the finding of an elevated red cell mass (44.6 ml/kg) and a normal plasma volume (44.8 ml/kg). The bone marrow aspirate showed myeloid hyperplasia with an increased number of megakaryocytes. The neutrophil alkaline phosphatase score was elevated (78). Oxygen saturation was normal. The erythropoietin level was markedly elevated at 0.63 iu/ml (normal 0.1-0.6 iu/ml).

Intravenous urogram showed a small left kidney with delayed excretion of contrast. Renal scan and renogram also showed impaired function on the left side. In view of these radiological abnormalities and the clinical finding of an epigastric bruit, an arteriogram was performed (Fig. 1). This demonstrated a severe stenosis of the left renal artery and moderate stenosis of the origin of the right renal artery. As well, a large collateral vessel ran from the region of the inferior mesenteric artery and supplied an occluded superior mesenteric artery (marginal artery of Drummond). Renal vein renin concentrations taken simultaneously were elevated: left renal vein 14.5 ng/ml/hr; right renal vein 6.8 ng/ml/hr; inferior vena cava 6.8 ng/ml/hr.

The patient was venesectioned on 4 occasions and then treated with radioactive phosphorous. His blood pressure responded to treatment with the angiotensin I converting enzyme inhibitor, captopril. A left nephrectomy was subsequently performed. He now has a normal blood pressure and does not require anti-hypertensive medication.

Discussion

A diagnosis of polycythaemia rubra vera was made on the findings of splenomegaly, an elevated white cell and platelet count with compatible changes in

FIG. 1. Bilateral renal artery stenosis and 'marginal artery of Drummond' (arrowed).

the bone marrow and normal oxygen saturation. Patients with this disease are often hypertensive although the reason is unclear. It is not related to blood viscosity (Lawrence, Berlin and Huff, 1953) and does not appear to resolve after venesection (Lawrence, 1949).

We have shown that, in our patient, hypertension was secondary to an ischaemic kidney. Significant renal artery stenosis was confirmed by the elevated renin levels and the demonstration of marked narrowing of both renal arteries on arteriogram. This is perhaps not surprising as occlusive vascular disease often occurs in conjunction with polycythaemia rubra vera. Cerebral, cardiac and mesenteric arteries seem to be particularly involved. To our knowledge, however, this is the first report of renal artery stenosis occurring in a patient with this disease. The unusual finding of an elevated erythropoietin level implies that the renal ischaemia was indeed marked and may even have led to an exacerbation of polycythaemia.

We suggest that renal arterial disease may have been overlooked in patients with polycythaemia rubra vera. The finding of refractory hypertension should arouse suspicion of this complication.

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