Severe hypertension and raised haematocrit: unusual presentation of Guillain-Barré syndrome

A.M. Richards, M.G. Nicholls, M.E.J. Beard, P.J. Parkin and E.A. Espiner

Departments of Endocrinology, Cardiology, Haematology and Neurology, The Princess Margaret and Christchurch Hospitals, Christchurch, New Zealand.

Summary: A 36 year old man presented with headache, polyuria, thirst and weakness of the lower limbs. Hypertension and a high haematocrit were striking features on initial assessment. Subsequently the full picture of Guillain-Barré syndrome developed. Hormone measurement revealed marked sympathetic nervous system and renin-angiotensin system activation with a return to normal of both blood pressure and hormones with neurological recovery.

Introduction

Hypertension has long been recognized as a possible complication of the Guillain-Barré syndrome (Haymaker and Kernohan, 1949). The elevation of blood pressure typically occurs after neurological signs are well-established. We report a patient who at clinical presentation had severe hypertension and a raised haematocrit.

Case report

A 36 year old upholsterer was admitted to hospital following 5 days of thirst, weight loss, headache, lower limb weakness, nocturia and frequency of micturition. Three years previously, mild hypertension had been noted (150/100 mm Hg) and subsequent typical blood pressure readings while taking amiloride and hydrochlorothiazide as 'Moduretic' daily, ranged from 140/90 mm Hg to 150/100 mm Hg. Examination revealed a debilitated man with a tachycardia (120 beats/minute) and a supine blood pressure of 200/175 mm Hg. The optic fundi were normal and apart from a generalized reduction in the deep tendon reflexes, no other abnormality was found. Investigations showed an elevated haemoglobin (19.8 g/dl) and haematocrit (61%), proteinuria and microscopic haematuria, but normal plasma urea, creatinine, and electrolytes, chest

X-ray and electrocardiogram. The combination of tachycardia, severe hypertension, and high haematocrit suggested the possibility of pheochromocytoma. Hence, oral labetalol was commenced after obtaining venous blood for hormone measurements. In view of elevated plasma catecholamines (Table I), an abdominal computed tomographic (CT) scan was performed but revealed normal adrenal morphology. Venesection (200 ml daily for 3 days) was carried out because of the raised haematocrit. However, blood volume studies on day 3, using ^{51}Cr -labelled red cells and ^{125}I -albumin, revealed a reduced total blood volume of 3.91 (predicted 4.51, 4.5 litres, i.e. 71.4 ml/kg). Red cell mass was slightly reduced at 1.32 litres. Venesections were stopped. Supine hypertension remained pronounced (175/150 mm Hg) but postural hypotension was striking (95/77 mm Hg standing). Weakness progressed and over 5 days he developed facial diplegia, paralysis of the jaw, palate and tongue and flaccid areflexic paralysis of all limbs. There was mild sensory impairment in the fingers and toes. Nasogastric feeding was needed but ventilatory assistance was not required. Cerebrospinal fluid protein was elevated (4.64 g/l) but the white cell count was normal. Hypertension persisted despite increasing doses of labetalol (1200 mg daily) and the addition of prazosin. Blood pressure control was achieved after prazosin was replaced by clonidine 150 μg thrice daily (Table I). After 1 week he began to show spontaneous improvement proceeding to full neurological recovery within 11 weeks. The supine blood pressure on the above medication was 140/90 mm Hg 4 months after presentation.

Special investigations showed activation of the sympathetic system (reflected by high plasma catecholamines) and the renin-aldosterone system,

A.M. Richards, M.B., Ch.B., M.R.A.C.P.; M.G. Nicholls, M.D., F.R.A.C.P.; M.E.J. Beard, F.R.A.C.P., F.R.C.P.E., F.R.C. Path.; P.J. Parkin, M.D., F.R.A.C.P.; E.A. Espiner, M.D., F.R.A.C.P.

Correspondence: A.M. Richards, Endocrinology Department, The Princess Margaret Hospital, Christchurch, New Zealand.

Accepted: 27 October 1983

Table I Laboratory and clinical details

Day in hospital	1	2	3	4	5	8	32	35	40
Medications		Labetalol					Prazosin		
							Clonidine		
Supine arterial pressure (mm Hg)	200/175	175/150			170/110		160/110		120/80
Haemoglobin (14–18g/dl)	19.8	20.1	18.3	15.6	14.4	14.5		13.6	
Haematocrit (40–52%)	61	60	53	45	44	43		41	
Plasma noradrenaline (100–800 pg/ml)	1723					983	284		
Plasma adrenaline (25–150 pg/ml)	314					407	95		
Plasma renin Activity (0.15–1.55 nmol/l/h)	10.3					4.3	2.0		
Plasma aldosterone (100–550 pmol/l)	1396					1023	207		
Plasma sodium (136–146 mmol/l)	135	128	127	130	133	136		138	
Plasma cortisol (110–570 nmol/l)	1030					918	170		

Normal control levels are given in parentheses

together with elevated plasma cortisol (Table I). With the exception of plasma renin activity, these indices were normal by the 32nd day. Plasma sodium concentration was initially normal, then declined over the next 2 days. Urine osmolality exceeded that in plasma at this time suggesting the possibility of high ADH levels. Follow-up urinalysis and intravenous urogram were normal.

Discussion

Severe hypertension associated with a raised haematocrit and developing at the same time or even before, neurological signs, has not previously been reported in the Guillain-Barré syndrome. The pathophysiological basis for hypertension in the Guillain-Barré syndrome is unclear and reports have variously implicated sympathetic overactivity (Mitchell and Meilman, 1967), increased renin release (Stapleton, *et al.*, 1978), an associated glomerulonephritis (Rodriguez-Iturbe *et al.*, 1973), and baro-receptor dysfunction (Tuck and McLeod, 1981). Hypervolaemic hypertension seems unlikely as our data suggest the blood volume was low, and venesection did not reduce supine pressures. Mild essential hypertension in our patient may have predisposed him to the striking blood pressure abnormality.

Whatever the underlying mechanism, we surmise

that blood pressure rose rapidly causing a 'pressure natriuresis' and thence plasma volume depletion with concomitant elevation of the haematocrit. Activation of the sympathetic and renin-aldosterone systems, and augmented release of ADH and ACTH presumably followed, or were further stimulated by, the fall in blood volume. In such circumstances blood volume studies were decisive in distinguishing relative from true polycythaemia. Hypertension plus a raised haematocrit has hitherto suggested renal or renal artery disease, pheochromocytoma, primary aldosteronism, polycythaemia rubra vera, and Gaisbock's syndrome ('stress polycythaemia'). Guillain-Barré syndrome may possibly now be added to this list.

Addendum

Since submission of this report Fagius and Wallin (1983) have reported sympathetic hyperactivity in 3 patients (with Guillain-Barré syndrome complicated by hypertension) assessed with microelectrode recordings of muscle nerve sympathetic activity. Sympathetic activity returned to normal with recovery.

Acknowledgements

We would like to thank Miss H. Legge and Miss D. Roberts who performed hormone assays. Mrs N. Purdue kindly typed the manuscript.

References

- FAGIUS, J. & WALLIN, G. (1983). Microneurographic evidence of excessive outflow in the Guillain-Barré syndrome. *Brain*, **106**, 589.
- HAYMAKER, W. & KERNOHAN, J.W. (1949). The Landry-Guillain-Barré syndrome. *Medicine (Baltimore)*, **28**, 59.
- MITCHELL, P.L. & MEILMAN, E. (1967). The mechanism of hypertension in the Guillain-Barré syndrome. *American Journal of Medicine*, **42**, 986.
- RODRIGUEZ-ITURBE, B., GARCIA, R., RUBIO, L., ZABALA, J., MOROS, G. & TORRES, R. (1973). Acute glomerulonephritis in the Guillain-Barré-Strohl syndrome. *Annals of Internal Medicine*, **78**, 391.
- STAPLETON, F.B., SKOGLUND, R.R. & DAGGETT, R.B. (1978). Hypertension associated with the Guillain-Barré syndrome. *Pediatrics*, **62**, 588.
- TUCK, R.R. & MCLEOD, J.G. (1981). Autonomic dysfunction in Guillain-Barré syndrome. *Journal of Neurology, Neurosurgery and Psychiatry*, **44**, 983.