Noninvasive haemodynamic studies in haemodynamically challenged patients
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Factitious phaeochromocytoma
CHAPTER 8

Introduction

A phaeochromocytoma, although often suspected in the case of severe paroxysmal hypertension, is seldom diagnosed. When a doctor suspects a phaeochromocytoma, an array of diagnostic tests can be performed, mostly without success. Various other diagnoses must be considered when a phaeochromocytoma cannot be diagnosed, but in approximately half of the patients with paroxysmal hypertension a definite diagnosis cannot be made. We describe the case of a woman with severe paroxysmal hypertension caused by Valsalva's manoeuvre during blood pressure measurement.

Case report

A 48-year-old woman was transferred to our hospital for the evaluation of severe therapy-resistant hypertension. Three months before admission to our hospital she was admitted to another hospital because of nausea, headache, palpitations and near-syncope, with a blood pressure of 250/160 mmHg while taking metoprolol. Her history revealed a borderline personality disorder; she had been admitted to psychiatric hospitals several times in the past 20 years because of suicide attempts and depressive episodes. She was wheelchair-bound despite the absence of an objective neurological disorder. Funduscopy showed grade II hypertensive retinopathy. She was transferred to our hospital because extensive workup could not demonstrate hormonal or renovascular hypertension and several antihypertensive agents, taken under supervision of a nurse, did not bring about a meaningful drop in pressure.

On admission, she complained of episodes of headaches, visual disturbances (diminished acuity at close range and flashes), anxiety, palpitations, nausea and occasional vomiting, lasting between several minutes and half an hour and occurring several times per day. Physical examination showed a nervous, wheelchair-bound woman. Cuff blood pressure varied between 95/60 mmHg and 240/170 mmHg, and her pulse rate was 80 to 165 bpm. She had scars of self-inflicted wounds on the backs of both hands, but otherwise physical examination was unremarkable. Driven by the suggestive complaints that were accompanied by a highly variable blood pressure and heart rate, further investigations to rule out a phaeochromocytoma were performed. Catecholamines were repeatedly normal both in urine and plasma. During an attack plasma noradrenaline rose to 4.66 mmol/L (normal values < 3.25 mmol/L) and adrenaline was 2.85 mmol/L (normal < 0.55 mmol/L). The clonidine suppression test was negative, as well as an octreotide scan. To investigate short-term blood pressure variability beat-to-beat finger blood pressure monitoring with Finapres Model 5 (TNO-BMI, Amsterdam, The Netherlands) on the left hand was performed. We observed repeated swings in blood pressure that typically accompany the various phases of Valsalva's
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manoeuvre. These episodes lasted well over 30 seconds. We performed simultaneous airflow measurement with a nose thermistor, CO$_2$ measurement in the exhaled air using a capnograph, and blood pressure measured on the contralateral arm using a Dinamap recorder (GE Medical Systems, Milwaukee, Wisconsin). The tracings showed that the patient performed Valsalva’s during each Dinamap measurement (see Figure, left) but also repeatedly while alone and unobserved. Typical Dinamap readings during Valsalva’s manoeuvre were 220/120 mmHg. These blood pressure responses were identical to those evoked when the patient was instructed to do a classic Valsalva’s manoeuvre with a pressure of 20 mmHg (Figure, right). When confronted with these findings, the patient said that she did not do it on purpose. She was referred to a psychologist specialized in breathing technique.

Figure 8.1 Blood pressure, heart rate (Finapres), airflow through the nose (nose thermistor) and CO$_2$ content of the exhaled air through the nose (capnography) during a Dinamap measurement (left) and instructed Valsalva’s manoeuvre (right). The arrow indicates elevated CO$_2$ level after resumption of breathing. *Erroneous lack of CO$_2$ due to predominant mouthbreathing.
Discussion

Severe, paroxysmal hypertension always suggests a phaeochromocytoma. When such a tumour cannot be diagnosed, which is often the case, several other diagnoses must be considered. Kuchel described a series of 63 patients [1] with complaints suggestive of phaeochromocytoma in whom the diagnosis could not be made, a so-called pseudopheochromocytoma. Of these patients 49 had hypertension and 8 patients were found to have both hypertension and hypotension. Diagnoses that were found included idiopathic hypovolemia (14 patients), mastocytosis (9 patients), incidentaloma (9 patients), neurogenic hypertension (4 patients) and cocaine abuse (1 patient). In our patient these diagnoses could be rejected. In the patients without an alternative diagnosis, a possible psychosomatic response to undetermined stimuli was considered. In a recent review article, [2] Mann described a series of 21 patients with paroxysmal hypertension in whom phaeochromocytoma could not be diagnosed. These patients were selected from 700 consecutive patients referred for the evaluation of hypertension. Although a relation to emotional distress was initially not apparent, later in the diagnostic course 14 patients acknowledged a history of severe emotional trauma. A rare diagnosis that falls within the same category of psychosocial related disease, although not diagnosed in the series of Kuchel [1] and Mann [2], is factitious phaeochromocytoma. According to the Diagnostic and Statistical of Mental Disorders, [3] patients with factitious disorder are fully aware of their actions, and produce or feign physical or psychological symptoms in order to assume the sick role. This is not done for economic gain or to avoid responsibility, such as in malingering. Clues leading to suspicion of factitious disorder are equanimity to diagnostic or therapeutic procedures, borderline personality traits, evidence of self-induced physical signs, deprivation in childhood and multiple hospitalisations, which were all present in our patient. Other clues are knowledge of, or experience in a medical field, antisocial personality traits, multiple scars (abdominal), male gender and a police record. Several case-reports describe patients who surreptitiously inject catecholamines. [4-6] Our patient, however, performed frequent Valsalva manoeuvres, elicited by blood pressure measurement, thereby creating a disproportionate high reading of her blood pressure. As such, the diagnosis of factitious disorder, or Munchausen syndrome could be made. This specific form of factitious disorder has been described once before. [7] Several similarities exist between that case and our patient. They also found a discrepancy between continuous measured blood pressure and blood pressures measured with a cuff. Furthermore, blood pressure control was unsuccessfully attempted with several anti-hypertensive agents, including intravenous medication. Further studies to investigate haemodynamic changes evoked by prolonged
straining in 3 healthy volunteers were performed by Kaisalam et al. [7]. The average rise in mean arterial pressure was 20 mmHg, with one patient attaining a blood pressure of 235/115 mmHg. We were able to reproduce the exact haemodynamic changes that were witnessed during blood pressure measurement, by instructing the patient to maintain an expiratory pressure of 20 mmHg, during 30 s.

In conclusion, we found in a hypertensive patient with suspected phaeochromocytoma severe hypertensive periods, caused by frequent voluntary Valsalva manoeuvres, particularly triggered by blood pressure measurements. Continuous noninvasive finger arterial pressure monitoring led to the recognition of the Valsalva's manoeuvre.
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References


