Pheochromocytoma or Münchausen syndrome: the masquerade is up

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Pheochromocytoma is a rare, deceptive and potentially deadly catecholamine-producing tumor, the recognition of which is often obscured and delayed by other more obvious diagnoses [1]. Diagnosis of pheochromocytoma is, however, also likened to finding a needle in a haystack so that most frequently when the tumor is suspected, the manifestations reflect an entirely different clinical condition. Differential diagnosis is made particularly problematic by the highly variable presentation of the tumor and difficulties in interpretation of biochemical tests of catecholamine excess, which can often return positive results due to activation of sympathoneural and adrenal medullary systems or other factors influencing measured levels of catecholamines and metabolites.

In this issue, Norcliff-Kaufmann et al. [2] report on a patient presenting with recurrent ventricular tachyarrhythmias resulting from surreptitious self-injection of epinephrine associated with Münchausen syndrome, a rare psychiatric disorder in which patients repeatedly seek medical attention for a factitious illness. This particular factitious disorder is characterized by a chronic and unremitting form of extreme behavior in which the individual affects dangerous manipulations of his or her own physical condition to simulate or create disease or injury, and in which the medical deceptions provide gratification from assuming the sick role. Besides the presently described patient, there have been at least seven other reports of patients who have feigned pheochromocytoma through self-administration of epinephrine or other manipulations

designed to simulate the tumor [3–9], indicating a need for more general consideration of the psychiatric disorder in the differential diagnosis of catecholamine-producing tumors.

The medical background of patients with Münchausen syndrome, typically associated with visits to numerous clinicians and specialists and multiple hospitalizations, is not that different from many patients in whom an autonomic disorder or pheochromocytoma is eventually considered. In the present case, the presentation of hypertensive episodes and ventricular tachyarrhythmias prompted numerous investigative and pharmacological interventions as well as more extreme surgical approaches, including ablation of the AV junction and implantation a dual chamber defibrillator. Associated issues of iatrogenic morbidity, patient confidentiality, qualifications for reimbursement of health care costs by insurance providers and accompanying medical-legal complications are all problems that can confuse and delay diagnosis of the underlying psychiatric condition. In such situations, direct communications among clinicians who have dealt with the patient are more likely to reveal the condition than the patient's own verbal history or forwarded medical records. Inconsistencies in the stories provided by patients are typical of Münchausen syndrome, and in the present case provided one of several clues to diagnosis of the true condition.

A history of childhood neglect or abuse, evidence of previous psychiatric problems, lack of stable relationships and extensive knowledge of symptomatology and the working of hospital systems to which the patient is drawn represent other clues to the diagnosis. In the present case, the patient's occupation as an allergy nurse provided her with access to epinephrine, a similar situation to previous cases of epinephrine self-administration [4, 6, 9]. The ensuing production of many of the classic signs and

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symptoms of pheochromocytoma, compounded by positive results of biochemical testing, can be fairly convincing of the tumor. In this particular patient, however, epinephrine self-injection was apparent from the measurements of plasma catecholamines during autonomic testing when the patient experienced an “attack” and when plasma epinephrine was measured at over 25,000 pg/mL. While solitary elevations of epinephrine with normal levels of norepinephrine are possible in patients with adrenal pheochromocytoma, this secretory pattern is extremely rare. More importantly, the completely normal plasma concentrations of metanephrine that were measured earlier had already excluded diagnosis of an epinephrine-producing tumor.

This interpretation follows from improved understanding of catecholamine metabolism, which now makes it clear that the metanephrines are produced within pheochromocytoma tumor cells continuously and independently of catecholamine release. Thus, in patients with pheochromocytoma, while catecholamines may be normal between episodes, the O-methylated metabolites are invariably elevated. Therefore, previous recommendations that biochemical testing is preferably carried out during episodes when patients are symptomatic—as has also been suggested in one other case of factitious pheochromocytoma [7]—are no longer tenable. To minimize false positive results, blood for plasma metanephrines is best collected when patients are asymptomatic, preferably after 30 min of supine rest.

As also outlined in two other reports [3, 4], a pattern of extreme elevations of epinephrine but normal levels of metanephrine excludes pheochromocytoma, but is entirely consistent with epinephrine self-administration. Therefore,

with measurements of metanephrines, particularly plasma free metanephrines, the masquerade is up; it is no longer feasible for clinicians to be deceived into a diagnosis of pheochromocytoma by a patient surreptitiously self-injecting epinephrine.

References

1. Manger WM (2009) The protean manifestations of pheochromocytoma. *Horm Metab Res* 41:658–663
2. Norcliff-Kaumann L, Gonzalez Duarte A, Martinez J, Kaufmann H (2010) Tachyarrhythmia with elevated cardiac enzymes in Münchausen syndrome *Clin Auton Res*
3. King TF, Shea PO, Sullivan EP, Srinivasan R, Griffin A, Fitzgerald R, Tormey W, Smith D (2008) An apparent pheochromocytoma and abnormal thyroid function tests. *Ann Clin Biochem* 45:447–448
4. Chidake AR, Pacak K, Eisenhofer G, Lawrence JE, Ayala AR (2006) Utility of plasma free metanephrines in diagnosis of factitious pheochromocytoma. *Endocr Pract* 12:568–571
5. Boon D, Kraaijenhagen RA, van Montfrans GA (2001) Factitious pheochromocytoma-like hypertensive bouts caused by the Valsalva maneuver. *Ann Intern Med* 135:304–305
6. Spitzer D, Bongartz D, Ittel TH, Sieberth HG (1998) Simulation of a pheochromocytoma—Munchausen syndrome. *Eur J Med Res* 3:549–553
7. Stern TA, Cremens MC (1998) Factitious pheochromocytoma. One patient history and literature review. *Psychosomatics* 39:283–287
8. Kailasam MT, Parmer RJ, Stone RA, Shankel S, Kennedy BP, Ziegler MG, O'Connor DT (1995) Factitious pheochromocytoma: novel mimicry by Valsalva maneuver and clues to diagnosis. *Am J Hypertens* 8:651–655
9. Keiser HR (1991) Surreptitious self-administration of epinephrine resulting in ‘pheochromocytoma’. *JAMA* 266:1553–1555