

Nephroquiz (Section Editor: M. G. Zeier)

A case of strange cardiac rhythms

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Background

Nephrologists must be good all-round clinicians. They should know a lot about primary and secondary hypertension, even if blood pressure is not in the foreground and the presentations are not as generally expected. Our patient presented intubated and on extracorporeal membrane oxygenation. Nonetheless, we got to the source of his problem.

Case

A 60-year-old professor of astrophysics falls from a ladder while picking cherries from the tree. An ambulance is called to take him to the hospital. Initially, he is conscious but confused. While in the ambulance, he develops supraventricular and ventricular arrhythmias and then ventricular fibrillation. He is resuscitated but unfortunately aspirates during intubation. No history is obtainable, but his wife reports that he was always healthy. In the emergency department, he required FiO₂ of 100%, positive end-expiratory pressures of >20 mmHg, and shortly thereafter, extracorporeal membrane oxygenation (ECMO). A routinely performed computed tomography (CT) scan showed no evidence of fractures, acute respiratory distress syndrome (ARDS) or an abdominal mass (Figure 1).

The 5 cm diameter cystic lesion residing adjacent to the left kidney and the 1.7 cm solid lesion found adjacent to the right kidney led us to perform some more diagnostic tests. The levels of plasma metanephrines were 1774 pg/mL (normal <80 pg/mL), and normetanephrines were 2471 pg/mL (normal <200 pg/mL). In the urine, epinephrine, norepinephrine, vanil mandelic acid and dopamine levels were all markedly elevated, while the serum aldosterone concentration was within the normal limits. Although the patient was not severely hypertensive in the

intensive care unit, we treated him preoperatively with 0.9% saline, phenoxybenzamine and then added nebivolol as labetalol is no longer available in Germany. Happily, the good professor recovered from ARDS, was stabilized and extubated. Cooperative surgeons removed the catecholamine-producing left adrenal mass laparoscopically after the additional imaging studies shown below were completed (Figure 2). The phaeochromocytoma was benign and the patient could leave the hospital much improved in all respects.

Discussion

Phaeochromocytomas are not that common, but usually turn up unexpectedly. They are catecholamine-producing neuroendocrine tumours arising from chromaffin cells of the adrenal medulla or extra-adrenal paraganglia. The tumours from extra-adrenal chromaffin tissue are referred to as extra-adrenal phaeochromocytomas or paragangliomas. The term paraganglioma is also used for the tumours derived from parasympathetic tissue in the head and neck, most of which do not produce catecholamines. Nearly 80–85% of phaeochromocytomas arise from the adrenal medulla, whereas about 15–20% are from extra-adrenal chromaffin tissue. The clinical presentation of phaeochromocytoma varies greatly, with similar signs and symptoms produced by many other clinical conditions [1].

During the past few years, a new heart disease syndrome has been recognized and precipitated by acute emotional or physical stress, now best termed as ‘stress cardiomyopathy’ (SCM) [2]. Exaggerated sympathetic stimulation may play a pathogenic role in the development of SCM. The plasma catecholamine levels have been found to be markedly elevated in some patients with SCM, and the syndrome has also been observed in other clinical states of catecholamine excess such as



Fig. 1. Would you care to make a diagnosis?

central neurologic injury and pheochromocytoma. Our patient had undergone cardiac compression during resuscitation and was critically ill, requiring ECMO, precluding a precise cardiac workup. We found no evidence of coronary disease and whether or not he had SCM prior to his fall is unknown. However, we believe that SCM played a role in his arrhythmias.

Our take-home message is to bear in mind the protean aspects of pheochromocytomas, another great masquerader. We counselled the good professor to leave the cherry picking to someone else.

Conflict of interest statement. None declared.

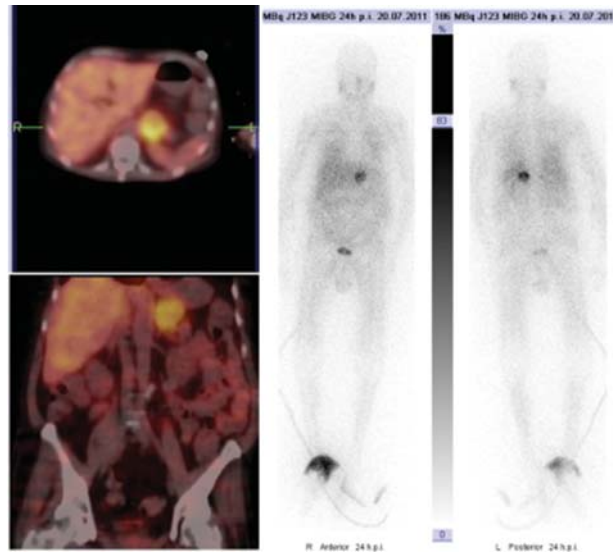


Fig. 2. Left panel shows CT positron emission tomography; right shows the results of meta-iodo-benzylguanidine (MIBG) scintigraphy (*Res ipse loquitur*).

References

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