A 42-year-old man was referred to our Hypertension Center for increasing episodes of tachycardia and elevated blood pressure (BP) readings. Over the past 3 years, he had been progressively experiencing frequent and worsening episodes characterized by palpitations, chest discomfort, and tremors, symptoms that the patient referred to as “a nervous heart.” These spells would only last a few minutes most of the time, sometimes completely resolving after 1 hour. On a few occasions, BP readings were elevated above 200/110 mm Hg. Multiple studies and tests performed at different medical centers, including plasma creatinine, thyroid hormones, aldosterone levels, rennin activity, and plasma fractionated metanephrines, were all normal. Full-body computed tomography scan and magnetic resonance imaging, renal artery duplex studies, electrocardiogram, transthoracic echocardiogram, and stress echo studies were also all unremarkable.

On presenting to our clinic, his resting heart rate (HR) was 62 beats per minute and BP was 118/70 mm Hg, without significant difference in all 4 limbs and without orthostasis. His physical examination, including a fundoscopic eye examination, was unremarkable. While examining his abdomen, the patient became tachycardic and diaphoretic, with frequent atrial and ventricular premature beats. His HR rose to 125 beats per minute and his BP to 190/102 mm Hg. Symptoms disappeared after 5 to 10 minutes. Plasma fractionated metanephrine levels were obtained that same day 2 hours after such an episode. Only metanephrine levels were slightly elevated at 80 pg/mL (normal <57 pg/mL) with normal normetanephrine values. Chromogranin-A levels were normal. The patient was sent home with an ambulatory HR and BP monitor. That night, the patient awoke from sleep again feeling palpitations and his heart pounding vigorously. HR and BP were elevated (Figure 1), both of which returned to normal values after <30 minutes.

A repeat computed tomography scan of the abdomen revealed normal-size adrenal glands. An iodine-131-metiodobenzylguanidine radioisotope scan was strikingly positive for isotope uptake from the left adrenal gland (Figure 2A and 2B). The patient was treated with the α-blocker phenoxybenzamine, 10 mg 3 times a day, and 1 week later, 20 mg of propranolol twice a day was added until both BP and HR were controlled. He then underwent laparoscopic surgery, and his left adrenal gland was successfully removed. The latter revealed the presence of adrenal medullary hyperplasia (Figure 3). The patient was discharged from the hospital 48 hours after the procedure and returned home the following week. Six months after the procedure, he remains normotensive and asymptomatic and on no medications.

Isolated adrenal medullary hyperplasia is an uncommon condition that can clinically mimic pheochromocytoma. Most commonly, the condition is associated with multiple endocrine neoplasia syndromes. Plasma metanephrine levels are normal or only slightly elevated, and imaging studies are negative for adrenal masses. It can affect 1 or both adrenal glands. The fact that, in this particular case, blood was drawn after the adrenal spell was precipitated (likely triggered by the physical examination) allowed us to detect the mild and often transient catecholamine levels that characterize these cases and therefore suspect the diagnosis. There is uncertainty whether this condition might antecede the formation of pheochromocytoma, and there have been cases also in which the contralateral gland medulla becomes affected after removing the affected one.

This condition represents a real challenge to the clinician, who might rule out pheochromocytoma in similar cases after a negative abdominal computed tomography scan or normal levels of serum biomarkers are obtained.

Disclosures

None.

References

Figure 2. A, Abdominal computed tomography scan (coronal view) showing normal adrenal glands. B, An iodine-131-metaiodobenzylguanidine scan revealing pronounced tracer uptake from the left adrenal gland (red arrow), suggesting hyperfunctioning adrenal gland medulla.

Figure 3. A, Low-power (hematoxylin and eosin stain, ×10 magnification) view revealing expanded adrenal gland medulla (arrowheads) surrounded by normal adrenal cortex (arrows). B, High-power view (hematoxylin and eosin stain, ×200 magnification) shows histologically unremarkable medulla. Findings are consistent with adrenal medullary hyperplasia.
A Nervous Heart
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