Adrenal incidentaloma that was not incidental: A case study

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Background

As a result of improved resolution of imaging, the finding of adrenal incidentalomas poses a fairly common clinical conundrum. Adrenal incidentalomas are defined as masses > 1 cm in diameter discovered accidentally on CT or MRI done for other reasons. CT scans alone reveal these masses in 4.4% of scans. Only about 5% of these masses are malignant, and only 10-15% are "functional," meaning that hormones are actively secreted. However, whenever an adrenal incidentaloma is identified, it is paramount that the mass is characterized with respect to: a) hormones secreted, if any; b) size and/or growth rate; and c) whether benign or malignant.

We report a case of an adrenal incidentaloma in a young adult that led to not one, but two diagnoses.

Description of a Case

A 20 year old woman presented to her primary care physician with symptoms of a urinary tract infection. The patient was also noted to have an elevation in blood pressure. To investigate the possibility of nephrolithiasis or pyelonephritis, a CT scan was performed. While no renal pathology was found, it did reveal the presence of a right adrenal mass. The mass was quite large, measuring 4.1 X 4.7 X 3.5 cm. Because of the this she was referred to an endocrinologist.

Endocrine evaluation

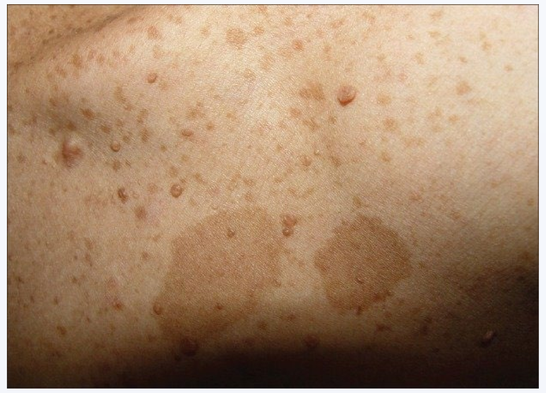
At that time, she admitted to frequent headaches, abdominal discomfort, palpitations, and tremor of her hands. She denied flushing and diaphoresis as well as nausea, vomiting, or diarrhea. Past medical history revealed that she had had no surgeries, or pregnancies. When asked about her BP she reported that her primary care physician had suggested she self-monitor it. Recent measurements were 157/92 and 160/117. She denied drinking, smoking, and drugs. On exam, she looked healthy. T 98.2, P 88, R 14, BP 152/102. Abnormal findings included > 10 café au lait spots on the abdomen, and several skin-colored or pink-tan papules and nodules on abdomen and back. The rest of the exam was unremarkable.

Laboratory Evaluation

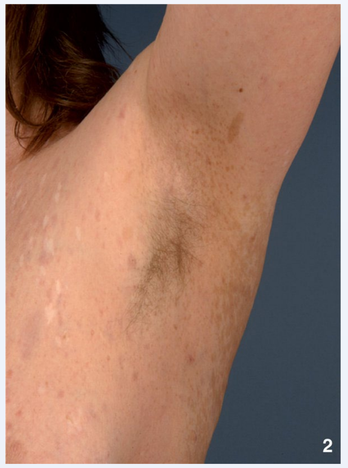
Complete blood count and comprehensive metabolic panel were both normal. Plasma free metanephrines She was placed on antihypertensive meds, and the following labs were obtained: CBC nl, metabolic panel nl. Plasma free metanephrines was 6.5 nmol/L (< 0.5). Plasma free normetanephrines was 1.9 nmol/L (< 0.9). Twenty-four hour urine studies revealed the following:

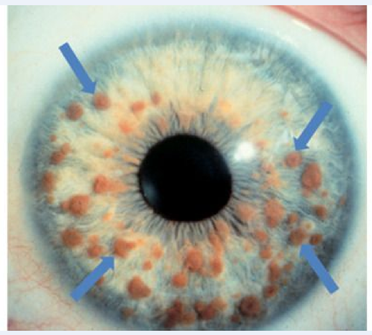
- Vanillylmandelic acid 21.3 mg (<7)
- Normetanephrines 6585 mcg (<900)
- Normetanephrines 1365 mcg (< 900)
- Total metanephrines 7951 (< 1300)
- Norepinephrine 264 mcg (< 80)
- Epinephrine 822 mcg (< 20)
- Dopamine 509 mcg (< 400).

Example of neurofibromas and café au lait spots



Example of axillary freckling and Lisch nodules of the iris





Adrenalectomy

Because laboratory evaluation indicated the presence of a pheochromocytoma, her antihypertensive medications were changed to the alpha blocker, phenoxybenzamine, followed by the beta blocker, metroprolol (after first ensuring that alpha blockade had been attained). Once titrated, this combination of antiadrenergic medications not only controls blood pressure but also protects these patients from the inadvertent tumor release of catecholamines during surgery.

Final Disposition

Postoperative follow up revealed the resolution of hyperadrenergic symptoms, as well as elevation of blood pressure. Pathological examination of the tumor was benign.

Referral to Genetics revealed not only the diagnosis of neurofibromatosis, type 1, but also noted the presence of both axillary and inguinal freckling as well as Lisch nodules.

Genetic testing revealed the specific mutation of the allele of the neurofibromatosis gene: 1466A>G (Y489C), which is a splicing and missense error.