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INTRODUCTION

Pheochromocytomas are chromaffin secreting tumors that arise from the adrenal glands or extra-adrenal sites. The usual symptoms in patients with pheochromocytoma include paroxysmal headaches, excessive sweating and tachycardia. Hypertension occurs frequently in patients with pheochromocytoma. About fifty percent of patients have paroxysmal hypertension and the rest have essential hypertension or normal blood pressure. Type 2 Diabetes or prediabetes also occurs in association with pheochromocytoma, although less frequently. The link between diabetes and pheochromocytoma is less well understood.

CASE PRESENTATION

We present a 30-year-old African American male with a five-year history of hypertension and later diagnosis of type 2 diabetes. His hypertension was suboptimally controlled, while taking four antihypertensive medications: Hydrochlorothiazide, Amlodipine, Enalapril and Atenolol. His type 2 diabetes was diagnosed approximately two years after an established history of hypertension. His diabetes was also suboptimally controlled on a single oral antidiabetic medication – Metformin. His A1C two months prior to presentation was 8.9%. There was no significant family history of diabetes or other endocrinopathies.

The patient was initially seen at the urology clinic for evaluation of microscopic hematuria. Further questioning revealed rather frequent episodes of palpitations, excessive sweating and generalized malaise. He denied bothersome headaches. Clinical examination showed a healthy appearing young man with a body mass index of 24.9. His resting heart rate was 104 beats per minute and sitting blood pressure was 199/103 mm Hg. The rest of his physical examination was unremarkable.

A renal ultrasound obtained at an outside facility depicted a 15 cm mass in the right renal angle. A subsequent CT urogram showed a 13.5 x 10.7 x 13.2 cm mass replacing the right adrenal gland with a central area of necrosis, calcification, and increased vascularity. These findings increased an earlier suspicion of a functional adrenal tumor and further investigation was pursued. His urine total catecholamine level was markedly elevated at 1659 (26–121) mcg/24 hrs, as was his urine total metanephrines 111,838 (90–630) mcg/24 hrs. His urine free cortisol was normal at 34 (4–50) mcg/24 hrs. The serum levels of aldosterone, renin and thyroid stimulating hormone were normal. The patient was appropriately prepped for surgery and underwent a successful right adrenalectomy three weeks after presentation. Postoperative pathology findings confirmed an 18.5 x 14.3 x 9.2 cm mass with features consistent with pheochromocytoma.

By his two-week postoperative clinic visit the patient had achieved optimal glycemic control without antidiabetic medications. His random blood glucose levels ranged from 80–145 mg/dL. In addition, his blood pressure was well controlled without any antihypertensive agent.

DISCUSSION

Both insulin resistance and decreased β-cell function have been postulated as mechanisms for hyperglycemia in patients with pheochromocytoma. β-adrenergic receptors mediate increased gluconeogenesis, glycogenolysis and decreased skeletal and adipose tissue glucose uptake. Wiesner and colleagues showed improved insu-
lin sensitivity in patients with type 2 diabetes-associated pheochromocytoma who underwent adrenalectomy. On the other hand, α-2 adrenergic receptors stimulation leads to decreased insulin secretion from β-cells. Glucose tolerance has been shown to improve with α-blockade alone in the absence of β-blockade.4

When they co-exist, early identification and treatment of pheochromocytoma in a patient with diabetes presents a rare opportunity to “kill two birds with one stone” in two otherwise difficult to manage conditions. Pheochromocytoma is one of the endocrinopathies associated with increased risk of diabetes in the group classified as “other specific types.”5 Other endocrinopathies that are associated with increased risk of diabetes include Cushing’s syndrome, hyperthyroidism, hyperaldosteronism, acromegaly, glucagonoma and somatostatinoma. While the reported incidence of diabetes in patients with pheochromocytoma is relatively low at 2–5%, the incidence of impaired glucose tolerance can be as high as 12–75%.1 In rare instances, patients with pheochromocytoma-associated diabetes have presented with diabetic ketoacidosis.6 Among 75 patients with pheochromocytoma reported in the series by Emmer et al, 12.5% of patients had impaired glucose tolerance while 2.8% had overt diabetes.7 Compared to those without diabetes, the patients with pheochromocytoma-associated diabetes tend to have a longer duration of hypertension, larger tumors and higher levels of urine catecholamine and metanephrine excretion.8 In general, glycemic control improves significantly after tumor excretion. Intermittent hormonal secretion and hypersecretion can develop in 20% of asymptomatic patients over time, thus repeat hormonal testing should be done annually for three to four years, even if the patient is asymptomatic.

**CONCLUSION**

This case illustrates the importance of screening for pheochromocytoma and other related endocrinopathies in young patients with diabetes and hypertension. When feasible, early diagnosis and treatment will significantly impact the patients’ outcome and quality of life.
REFERENCES


