Clinical Findings in Pheochromocytoma

Sir,

I have read the article by Khan *et al.* published in the 4th issue of 29th volume of Journal of the College of Physicians and Surgeons Pakistan with great interest.[1] Pheochromocytoma is an uncommon endocrine tumor, and a treatable cause of hypertension. Here, we aimed to focus on the clinical manifestations of pheochromocytoma, which can have a very wide clinical spectrum.

Pheochromocytoma is the name given to the catecholamine producing tumors of the adrenal medulla. They arise from chromaffin cells and usually occur in 4th or fifth decades; however, they can manifest at any age and in either gender. Incidence of pheochromocytoma is about 8 per million patient years and constitutes the underlying cause in two of every 1000 hypertension cases.[2] Aside from sporadic cases, Von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia type 2 (MEN 2), neurofibromatosis type 1(NF1) and paraganglioma syndromes are associated with pheochromocytoma.

The sign and symptoms of the disease are caused by hypersecretion of norepinephrine, epinephrine or dopamine. At presentation, about half of the cases with pheochromocytoma have paroxysmal symptoms. Tachycardia, headache and excessive sweating in episodes are classical symptoms of the disease. Pheochromocytoma is diagnosed in nearly half of the cases as adrenal incidentaloma in asymptomatic patients.

Elevated blood pressure, either episodic or constant, is the most common clinical finding in patients with pheochromocytoma. However, one of every 10 patients may have normal blood pressure, and even orthostatic hypotension may develop.[3] In particular, predominantly epinephrine-secreting pheochromocytoma cases may present with hypotension or even shock. Six and nine of every 10 symptomatic pheochromocytoma cases suffer from excessive sweating and headache, respectively.

Cardiomyopathy is a rare finding of the disease and caused by overproduction of catecholamines, which resembles Takotsubo cardiomyopathy.[4] Weight loss, polydipsia, polyuria, constipation, ocular signs including papill edema and blurred vision, may accompany pheochromocytoma.

Deteriorated carbohydrate metabolism may manifest as hyperinsulinemia, hyperglycemia or even overt diabetes mellitus.[5] Additional laboratory findings include leukocytosis, elevated erythrocyte sedimentation rate and secondary erythrocytosis.

Some patients suffer from elevated blood pressure, palpitations or arrhythmias during anesthesia, surgery or interventions (*e.g.* colonoscopy), after consuming tyramine containing foods or drinks, and by using medications (*e.g.* tricyclic antidepressants, corticosteroids, beta-adrenergic blockers, monoamine oxidase inhibitors).

In conclusion, since the clinical spectrum of the disease is very wide, clinicians should be alert about the signs and symptoms of patients with pheochromocytoma for the sake of diagnostic accuracy.

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Gulali Aktas

Department of Internal Medicine, Faculty of Medicine, Abant Izzet Baysal Universty, Bolu, Turkey

Correspondence to: Dr. Gulali Aktas, Department of Internal Medicine, Faculty of Medicine, Abant Izzet Baysal Universty, Bolu, Turkey
E-mail: draliaktas@yahoo.com

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AUTHOR'S REPLY

Sir,

It is true that pheochromocytoma has diverse clinical manifestations. Apart from sporadic cause, it may present as part of syndromes such as von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia type 2 (MEN 2), and neurofibromatosis type 2 (NF2). Therefore, screening to rule out associated anomalies has a pivotal role in the management of this disease. The most common and consistent symptom of pheochromocytoma is hypertention and about half of the cases present with intermittent hypertension.[1,2] Episodic hypertension at a young age of 17 years certainly raised suspicion of the presence of pheochromocytoma, which was then investigated thoroughly.

Vanillyl mandelic acid (VMA) level in 24-hour urine was raised and CT scan abdomen with intravenous contrast also supported the diagnosis. Although laproscopic adrenelectomy is the preferred treatment option; but in our patient, laparotomy was done for this purpose.

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Junaid Zia Hashmi

Department of General Surgery, Nishtar Hospital, Multan, Pakistan

Correspondence to: Dr. Junaid Zia Hashmi, Department of General Surgery, Nishtar Hospital, Multan, Pakistan
E-mail: juneehashmi@yahoo.com