Multiple retroperitoneal paragangliomas

Sir,

Pheochromocytomas and paragangliomas are rare neuroendocrine tumors arising from the adrenal medulla and extra-adrenal ganglia, respectively.

Retroperitoneal paragangliomas arise from paraganglia collections of specialized neural crest cells symmetrically distributed along the aorta in close association with the sympathetic chain.[1] In recent series, approximately one-third to one-half of paragangliomas are associated with an inherited syndrome.[2]

A 40-year-old male, with a medical history of type 2 diabetes and dyslipidemia, was referred to our department for paroxysmal hypertension.

At the age of 7 years, he was diagnosed with primitive polycythemia and treated with monthly phlebotomies. There was no family history of neuroendocrine tumor or polycythemia.

Since 6 months, the patient started to experience headaches, palpitations, and sweating. In addition, he presented a weight loss of 7 kg.

On examination, he had a body weight of 66 kg, a body mass index of 20.8 kg/m^2, and a blood pressure of 120/80 mmHg. There was no orthostatic hypotension. The

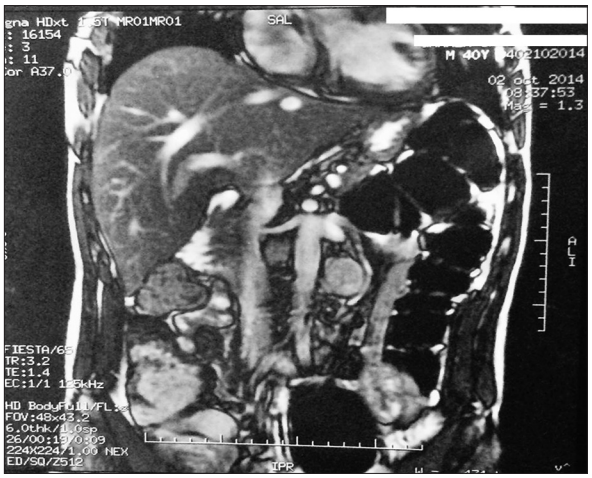

Figure 1: Preoperative magnetic resonance imaging showed multiple retroperitoneal tumors

rest of physical examination was normal. Electrocardiogram showed a sinus regular rythm (95/min).

24-h holter monitoring of blood pressure showed paroxysmal hypertension with peaks of 200/110 mmHg. Echocardiography revealed moderate hypertrophy of the left ventricle. Systolic and diastolic functions were preserved.

Laboratory tests demonstrated a high level of urinary normetanephrine (5495 nmol/24 h; reference range <213 nmol/24 h).

Computed tomography and magnetic resonance imaging of the abdominopelvic region revealed six retroperitoneal tumors measuring 11, 13, 27, 15, 40, and 37 mm, respectively [Figures 1]. Left and right adrenal glands were normal.

131-metaiodobenzylguanidine scintigram shows an intense radiotracer uptake by the six tumors.

The diagnosis of multiple retroperitoneal paragangliomas was established. After medical preparation, seven solid, encapsulated tumors were resected. Intra-operatively, four tumors were inter-aortico cave located and three tumors located below the left renal hilum. Histopathologic examination confirmed the diagnosis of multiple paragangliomas.

After surgery, his blood pressure was within normal range without medications and plasma level of noremetanephrine decreased to 153 nmol/24 h.

Paragangliomas are rare with multicentricity being more common in patients with familial history.[3] In fact, family history for paraganglial tumors, multiple tumors, and young age at diagnosis are associated with a high probability of finding a germline mutation.[4]

Our patient presented with a very rare association: Multiple paragangliomas and primitive polycythemia. Recently, a new syndrome of paraganglioma, somatostatinoma, and polycythemia has been discovered.[5]

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Conflicts of interest

There are no conflicts of interest.

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