Simultaneous Adrenal Pheochromocytoma and Carotid Body Paraganglioma in a Woman

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Simultaneous occurrence of carotid body tumor and pheochromocytoma is rare. Most pheochromocytomas have grown on adrenal medulla, but some of the pheochromocytoma patients have multifocal paragangliomas arising from extraadrenal tissues [1–3]. Pheochromocytomas and paragangliomas occur as sporadic tumors or they can be associated with several hereditary syndromes such as (1) multiple endocrine neoplasia type 2 (MEN 2), (2) Von-Hippel-Lindau disease (VHL) and (3) neurofibromatosis type 1 as an unusual genetic cause of pheochromocytomas. Genetic testing is recommended for patients with an apparently sporadic pheochromocytoma under the age

of 20 years with a family history or features suggestive of hereditary pheochromocytoma or for patients with sympathetic paragangliomas [4, 5]. For individuals who do not meet these criteria, genetic testing is optional. Discovery of pheochromocytoma or paraganglioma in a patient should lead to a careful search to rule out multifocal lesions and/or hereditary syndromes. The diagnosis of pheochromocytoma and paraganglioma is made by biochemical testing, and imaging is done to localize the tumor for surgical planning [6, 7]. F-18 FDG PET has proved to be an effective tool in the localization of pheochromocytomas and paragangliomas (Figs. 1 and 2).

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Fig. 1 A 45-year-old female was admitted to the hospital with uncontrolled hypertension. In laboratory analysis, urine norepinephrine was 26.0 $\mu\text{g}/24\text{ h}$ (normal, 12.1–85.5), epinephrine was 8.3 $\mu\text{g}/24\text{ h}$ (normal, 0.5–22.4) and normetanephrine was 568.1 $\mu\text{g}/24\text{ h}$ (normal, 88–444). F-18 FDG PET/CT was performed for further evaluation. PET/CT (a–c) showed intense focal FDG uptake (*arrowhead*) in the right adrenal gland. In addition, another intense focal FDG uptake (*arrow*) was noted in the right lateral neck. Enhanced neck CT (d) showed a 3-

cm-sized mass (*arrow*) with homogeneous enhancement at the level of the right carotid artery bifurcation. The mass splayed the carotid bifurcation and displaced the internal jugular vein. These are classic findings of carotid body tumor. Enhanced abdomen CT (e) revealed an intensely enhancing mass (*arrowhead*) in the right adrenal gland with few central non-enhancing areas suggestive of necrosis. These imaging features with abnormal laboratory finding were suggestive of pheochromocytoma

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Fig. 2 Surgical excision was performed for a right carotid mass and right adrenal mass. The microscopic findings of the adrenal mass (a) showed a typically organoid pattern ('Zellballen') of the normal paraganglion, which is composed of two cell types: chief cells with pale eosinophilic cytoplasm and slightly to moderately atypical nuclei and sustentacular cells. The microscopic findings of the carotid mass (b) showed the characteristic 'Zellballen' architecture, and a single pleomorphic cell was noted in the upper center. Therefore, adrenal pheochromocytoma and carotid paraganglioma were histologically confirmed