Correspondence

Pheochromocytoma: Another Neuroendocrine Tumor that Substantially Affects the Heart

To the Editor:

The February 2019 issue of the *Journal* included an excellent review of carcinoid heart disease (HD) by Ram and colleagues.[1] Practitioners may not be aware that pheochromocytoma, another neuroendocrine tumor, can also cause substantial cardiac injuries. There are important differences between cardiomyopathy in carcinoid HD and pheochromocytoma-induced cardiomyopathy.

Carcinoid HD usually involves only the tricuspid valve and right ventricle.[1] Pheochromocytoma manifests itself in a much wider spectrum of cardiac abnormalities, such as myocarditis, congestive heart failure, left ventricular thrombus, arrhythmia, and cardiac arrest; however, the cardiac valves are spared.[2-4]

Carcinoid HD is a late manifestation of carcinoid tumor after extensive liver metastasis; therefore, the prognosis is poor. In contrast, about 10% of pheochromocytomas initially present as severe cardiomyopathy in apparently healthy people, but without the classic symptom of hypertension and the paroxysmal symptoms of headache, diaphoresis, and palpitation. When diagnosed correctly, pheochromocytoma-induced cardiomyopathy is reversible after tumor removal, and the prognosis is favorable if it is properly managed.

The pathogenetic mechanism of carcinoid HD, predominantly serotonin-mediated, leads to tricuspid valve fibrosis and insufficiency. The more complex pathogenesis of pheochromocytoma-induced cardiomyopathy involves the release of catecholamines and other substances from the tumor. The only definitive histologic change is myocardial necrosis and fibrosis.[5,6]

Most cardiologists and oncologists are aware of the link between carcinoid HD and cardiomyopathy, so the cardiomyopathy is usually found during standard monitoring. Conversely, the link between pheochromocytoma and cardiomyopathy is not as well known, and diagnosis may be made only after computed tomograms to investigate pulmonary embolism or aortic dissection incidentally reveal a large adrenal mass. Therefore, treatment is often delayed.

Treatment of carcinoid HD involves reducing the tumor burden and surgically correcting tricuspid valve insufficiency. Treatment of pheochromocytoma-induced cardiomyopathy involves cardiac support, preoperative management, and tumor resection.

Pheochromocytoma is such a rare cause of cardiomyopathy that it may not be part of the initial differential diagnosis. After more typical causes are ruled out, it should be considered, especially when an adrenal mass is found incidentally.

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