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Robert Schreiner DO

Lehigh Valley Health Network, robert.schreiner@lvhn.org

Yehia Y. Mishriki MD

Lehigh Valley Health Network, Yehia.Mishriki@lvhn.org

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Malignant Paraganglioma-associated Takotsubo Cardiomyopathy

Robert S. Schreiner, DO and Yehia Mishriki, MD

Lehigh Valley Health Network, Allentown, Pennsylvania

Introduction

The WHO: Paragangliomas, uncommon catecholamine-producing tumors arising from extra-adrenal chromaffin cells. Catecholamines released from the tumor stimulate both alpha- and beta-adrenergic receptors.

The WHERE: Pheochromocytomas arise in the adrenal medulla. However, when catecholamine-producing tumors occur in extra-adrenal sympathetic ganglia, they are called paragangliomas. They may occur sporadically or as part of a hereditary syndrome (i.e. MEN2, von-Hippel Lindau syndrome).

The HOW: About 10% of paragangliomas are malignant often arising in the setting of germline mutations in a mitochondrial complex subunit-succinate dehydrogenase (SDH).

The WHAT: Malignant catecholamine stimulation can result in catastrophic effects including a stress cardiomyopathy (takotsubo cardiomyopathy), characterized by left ventricular dysfunction and potential cardiovascular collapse.

The WHY: Catecholamine-secreting tumors should be included in the differential diagnosis of takotsubo cardiomyopathy if other causes have been ruled out. If metastatic paragangliomas are found, genetic testing is warranted to seek out mutations, especially SDHB.

Case Presentation

A 54 year-old male presented with unexplained chest pain. An exercise stress test elicited frequent PVCs, ventricular tachycardia, and idioventricular rhythms. A subsequent cardiac catheterization showed mild luminal irregularities of coronaries; however, a left ventriculogram showed ejection fraction of 35% with evidence of apical ballooning consistent with a takotsubo cardiomyopathy. An evaluation coincidentally identified para-aortic and retroperitoneal masses. Circulating free catecholamines and 24-hour urinary metanephrines were elevated. He was diagnosed with a paraganglioma-like tumor. Following excision of his paragangliomas, LV size and function normalized. Beta blocker therapy was initiated with resolution of PVCs and improvement of symptoms. Molecular testing identified a succinate dehydrogenase type B mutation in the paraganglioma, a feature typically associated with malignancy.



Figure 1. Sagittal view and coronal view of thoracic and lumbar spines, showing left paravertebral mass at T7-T10 (encircled).



Figure 2. Coronal view, octreotide scan: White arrow consistent with an octreotide avid tumor left-sided paraspinous mass at the T9-T10 level.

Discussion

Paraganglioma-associated takotsubo cardiomyopathy has not been widely reported. This case **underscores the strong association between excess sympathetic stimulation and takotsubo contractile pattern.**

- Hypertension, headache, and diaphoresis suggest an early functional tumor. However, **takotsubo cardiomyopathy and cardiogenic shock may occur late** in an asymptomatic patient.
- If malignant paragangliomas are diagnosed, a **baseline cardiovascular evaluation should be initiated**, which should include electrocardiogram and echocardiogram in case subsequent cardiovascular collapse occurs.
- **Patients with paragangliomas should undergo genetic testing** to identify mutations associated with malignancy, most notably succinate dehydrogenase type B mutations. If malignant-associated mutations exist, close follow-up.
- **Catecholamine secreting tumors should be kept high on the differential** if a patient presents with takotsubo cardiomyopathy in the absence of an obvious trigger.

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