Malignant Paraganglioma-associated Takotsubo Cardiomyopathy

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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.

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.


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.