Diagnostic Imaging and Treatment of a Left Atrial Paraganglioma

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A 24-year-old woman presented for evaluation of episodic hypertension (blood pressure up to 190/122 mmHg) associated with palpitations. She used no illicit drugs. Holter monitoring revealed no arrhythmia. A 24-hour urine test revealed extremely high levels of catecholamine metabolites (vanillylmandelic acid, metanephrine, and normetanephrine). An echocardiogram showed a left atrial (LA) mass (Fig. 1). An iodine (I-123) metaiodobenzylguanidine (MIBG) single-photon-emission computed tomogram showed intense tracer uptake only in the subcarinal area (Fig. 2). A T1-weighted echo inversion-recovery cardiac magnetic resonance sequence revealed a nonmobile, well-circumscribed 2.4 × 3.2 × 2.7-cm mass in the posterior LA wall (Fig. 3). The mass had a high signal intensity on T1- and T2-weighted images, with substantial contrast enhancement.

To prevent a hypertensive crisis during surgery, we treated the patient with phenoxybenzamine for several days. The mass was in the LA roof, between the right and left pulmonary veins (Fig. 4). We resected the tumor with a margin of normal atrial tissue (Fig. 5) and closed the incision by using autologous pericardium. Histologic findings

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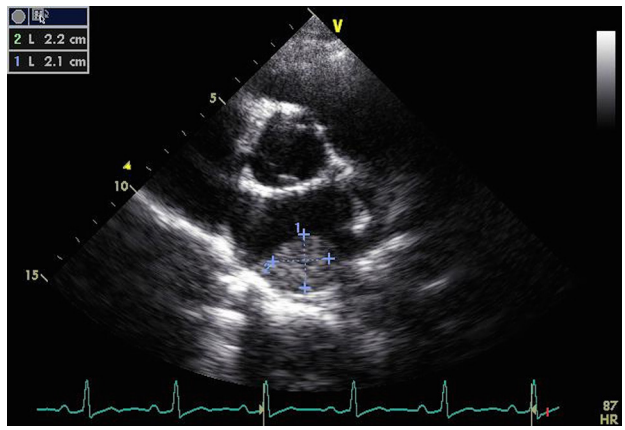

Fig. 1 *Transthoracic echocardiogram (parasternal short-axis view) shows the left atrial mass.*

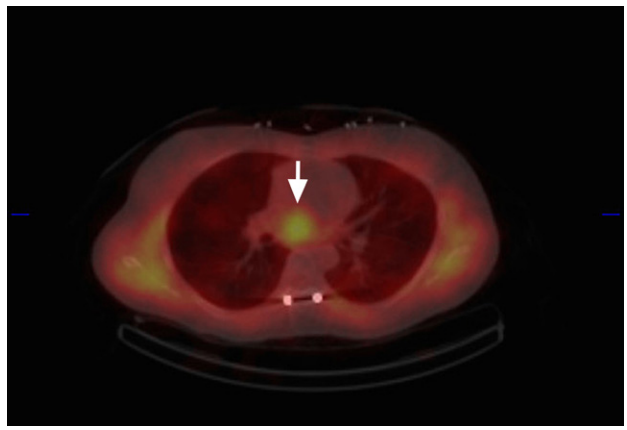

Fig. 2 *Single-photon-emission computed tomogram reveals intense I-123 metaiodobenzylguanidine uptake in the left atrium.*

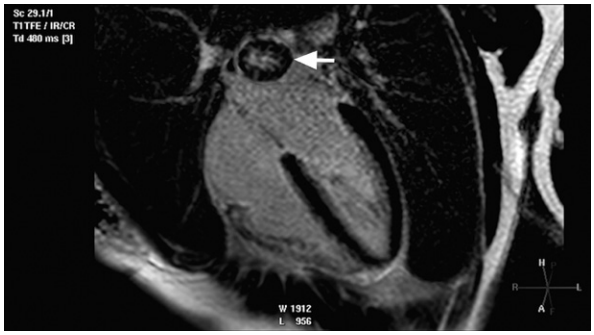

Fig. 3 *T1-weighted, echo inversion-recovery cardiac magnetic resonance sequence shows a well-demarcated mass (arrow) in the posterior wall of the left atrium.*

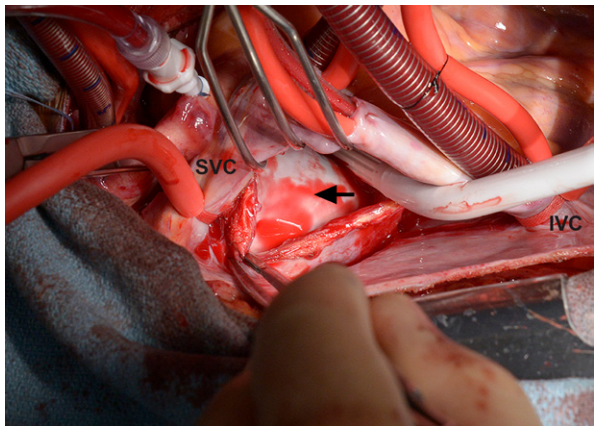

Fig. 4 *Intraoperative photograph shows a left atriotomy in the Sondergaard groove and cannulation of the superior vena cava (SVC) and inferior vena cava (IVC). The endocardial surface of the tumor bulges into the left atrium (arrow).*

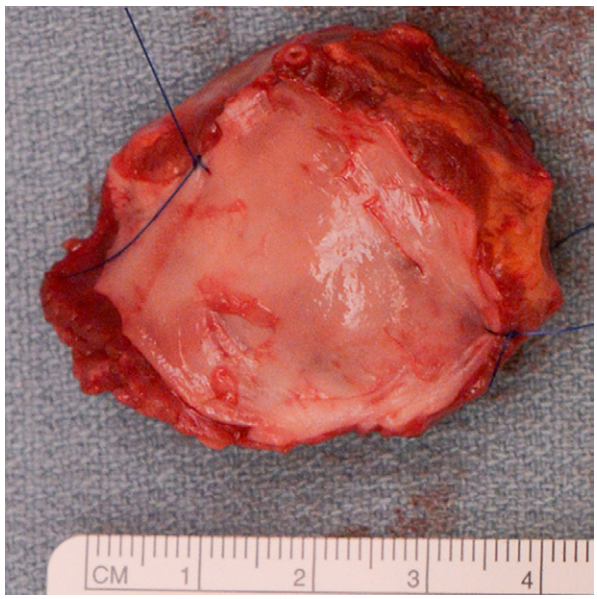

Fig. 5 *Photograph shows the endocardial surface of the resected mass.*

were consistent with a paraganglioma; there was no sign of invasion of surrounding tissues, suggesting that the tumor was benign (Fig. 6).[1] Two months later, the patient's symptoms had resolved and her catecholamine levels were normal.

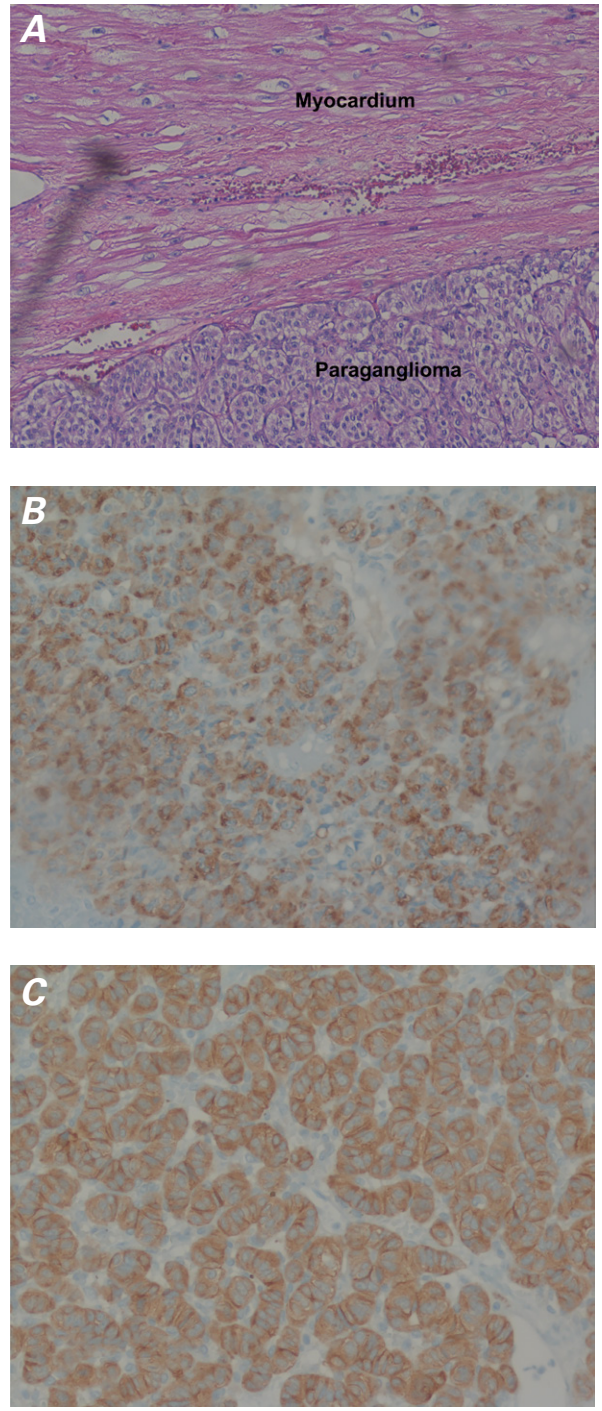

Fig. 6 ***A****) Photomicrograph reveals a distinct myocardial–paraganglioma border (H & E, orig. ×100). Chief cells also stained positive for* ***B****) chromogranin (orig. ×400) and* ***C****) synaptophysin (orig. ×400), consistent with pheochromocytoma of the chromaffin type.*

Comment

The prevalence of catecholamine-secreting tumors in hypertensive patients is <0.2%.[2] According to the most recent classification of tumors by the World Health Organization (2004),[3] paragangliomas develop from neural crest tissue in the sympathetic or parasympathetic system. The term pheochromocytoma is used for paragangliomas that arise from the adrenal medulla.[3] In 2 large series of cardiac paragangliomas,[4,5] 31% and 79% of tumors were hormonally active, as shown by high urine catecholamine levels.

High catecholamine levels are more sensitive than specific in diagnosing paragangliomas.[2] The molecular structure of MIBG is like that of norepinephrine and is avidly taken up by adrenergic tissues. We used radiolabeled MIBG scanning, which, unlike computed tomography, has a specificity of 100% for localizing paragangliomas.[6] We identified the anatomic relationship between the mass and the surrounding tissues by using cardiac magnetic resonance, which facilitated its resection. Careful testing and appropriate imaging enabled the early diagnosis and successful treatment of this hormonally active LA paraganglioma.

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