







Intramyocardial Hemangioma Mimicking Paraganglioma in Cardiac MRI and DOTANOC PET

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Indian J Radiol Imaging

Abstract

Keywords

- ▶ cardiac hemangioma
- ▶ cardiac paraganglioma
- ▶ DOTATATE PET

A cardiac hemangioma is a rare primary benign intracardiac tumor characterized by proliferation of endothelial cells. The lesions are hyperintense on T2/short tau inversion recovery magnetic resonance imaging (MRI) and show avid enhancement and retention of contrast agents. Here we present a case of a histopathologically proven cardiac hemangioma showing uptake on Dodecanetetraacetic acid - octreotate (DOTATATE), masquerading as paraganglioma in MRI and positron emission tomography imaging.

Case Presentation

A 42-year-old gentleman presented with multiple episodes of palpitation lasting for around 3 to 5 minutes. He had 15 to 20 self-limiting similar episodes in the last 6 months. None of the attacks were associated with syncope, chest pain, or dyspnea. The electrocardiogram showed narrow complex tachycardia with a right bundle branch block. Echocardiography revealed a well-defined encapsulated mass lesion in the left ventricular outflow tract without any outflow obstruction. Magnetic resonance imaging (MRI) to characterize the lesion showed a well-defined intramural lesion of measuring 5 × 4 cm in the anterior segment of the basal left ventricular myocardium. The lesion was isointense on T1-weighted imaging (T1WI) and avidly hyperintense on T2W sequences. There was intense enhancement in the arterial phases of the perfusion sequences, which persisted in the delayed venous phases (▶ **Fig. 1**). The native T1 and T2 values of the lesion were elevated (T1 value of 1,150–1,200 milli-

seconds and native T2 value of 60–70 milliseconds). Given avid T2/short tau inversion recovery hyperintensity and intense early as well as late enhancement, the differentials were of cardiac hemangioma and paraganglioma (PGL). Ga-68 DOTANOC positron emission tomography (PET) scan was performed to differentiate between the two. DOTANOC PET showed avid uptake and we zeroed down to a diagnosis of cardiac PGL (▶ **Fig. 2**). The urinary metanephrine and VanillylMandelic Acid (VMA) levels were however normal.

Since the patient was symptomatic of palpitations, the imaging features were suggestive of PGL and the lesion itself was well encapsulated, surgical resection was performed. On histopathology, the lesion was shown to have multiple slit-like vascular spaces. On immunohistochemistry, the lesion showed smooth muscle antigen and CD31 positivity (▶ **Fig. 3**), suggesting intracardiac hemangioma. The patient was discharged on postoperative day 5. Clinical follow-up after 2 years showed complete resolution of symptoms and no recurrence of the lesion in echocardiography.

DOI <https://doi.org/10.1055/s-0044-1800863>.
ISSN 0971-3026.

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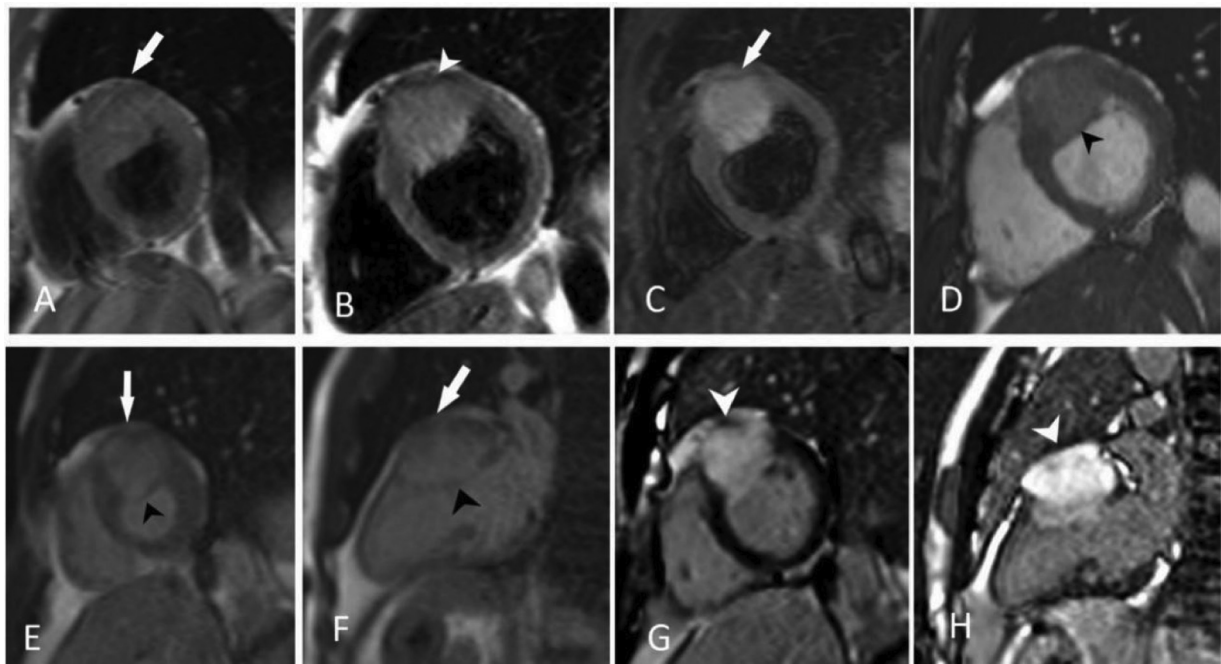


Fig. 1 An intramyocardial mass lesion (A) isointense to the myocardium in T1 image, (B) hyperintense on T2 image, and (C) short tau inversion recovery image. (D) Steady-state free precession sequence shows thin rim of normal myocardium (*arrowhead*), suggesting the lesion is intramural. (E, F) In perfusion imaging, the lesion shows intense enhancement in the first pass perfusion (*arrows*) with persistent enhancement in the (G, H) late gadolinium images.

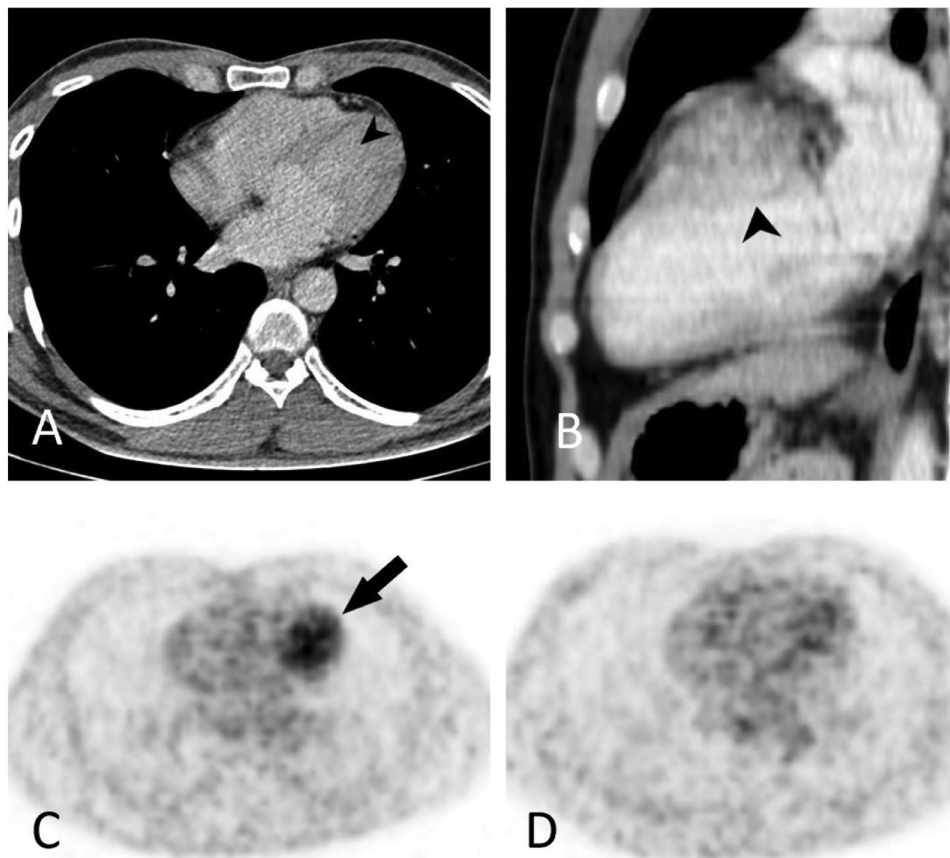


Fig. 2 (A) Axial and (B) oblique sagittal contrast-enhanced computed tomography (CECT) images show an enhancing lesion (*arrowhead*) near the Left Ventricular Outflow Tract (LVOT). (C) Ga-68 DOTANOC positron emission tomography CT shows uptake in the lesion (*arrow*) with (D) no uptake noted in the rest of the myocardium.

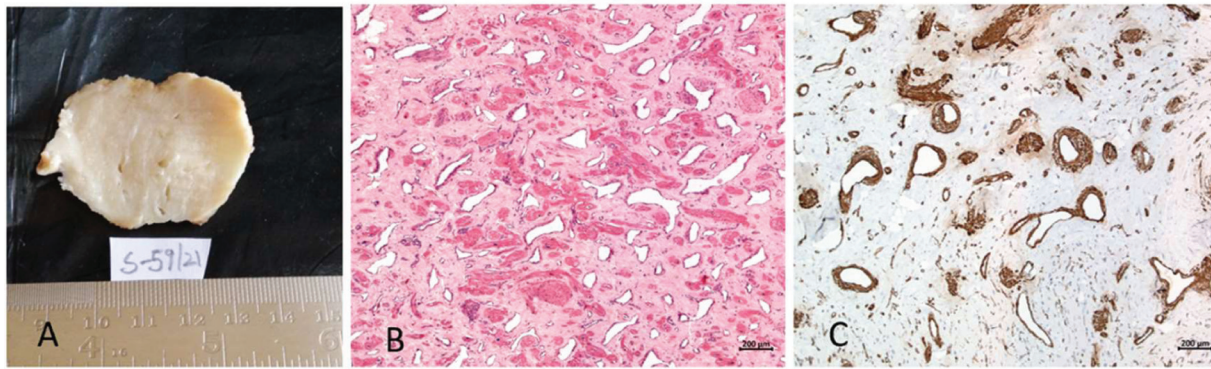


Fig. 3 (A) Gross pathological specimen of a lesion measuring 5 × 4 cm with (B) slit-like intramyocardial spaces on histology. (C) It is composed of arterioles and venules showing smooth muscle antigen positive in immunohistochemistry.

Discussion

The differential diagnosis of an intensely enhancing myocardial mass lesion can be cardiac metastasis of a highly vascular primary lesion, cardiac hemangioma, or cardiac PGL.^{1–3} Metastases can be ruled out by a screening contrast-enhanced computed tomography (CECT)/MRI scan of the chest and abdomen. Differentiation between hemangioma and PGL is of clinical importance.

PGLs are extra-adrenal neuroendocrine tumors (NETs) secreting adrenaline/noradrenaline or dopamine. These chemicals can lead to life-threatening arrhythmias and thus treatment of PGL is usually surgical resection.⁴

On the other hand, cardiac hemangiomas can be managed conservatively with a trial of beta-blockers or propranolol.⁵ Since PGLs are also secretory tumors, patients with PGLs are preoperatively treated with alpha receptor antagonists to prevent any intraoperative hypertensive crisis. Such preoperative preparations are not needed for cardiac hemangiomas.⁴

Thus, preoperative differentiation of PGLs and cardiac hemangiomas is of utmost importance, and Ga-68 DOTANOC PET is the usual investigation of choice. PGLs being NETs have somatostatin receptors and DOTANOC has a high affinity to these receptors. These PGLs are conventionally thought to be Ga-68 DOTANOC PET positive, while cardiac hemangiomas are negative.⁴

However, there are several previous case reports have described extracardiac vertebral or intraosseous hemangiomas showing uptake on DOTANOC PET. The cause of this uptake is likely the expression of somatostatin receptors in the endothelium of the blood vessels of hemangioma.⁶

To our knowledge, only one case of cardiac hemangioma showing DOTANOC uptake has been reported in the literature to date.⁷ The case reported by Shah et al was similar to ours; the preoperative provisional diagnosis was PGL because of DOTANOC avidity, but hemangioma was confirmed by histopathology.

Conclusion

To conclude, cardiac hemangiomas can mimic cardiac PGLs on both MRI and Ga-68 DOTANOC PET scan.

Authors' Contributions

J.V. conceptualized the study. A.R.C. and J.V. contributed to data collection and interpretation. A.R.C., J.V., and A.A. contributed to manuscript and figure preparation and manuscript revision based on feedback from co-authors. J.V. and A.A. supervised the study and made critical revision of the manuscript. All authors reviewed the results and approved the final version of the manuscript.

Patient Consent

The authors confirm that written consent for submission and publication of this case including images and associated text has been obtained from the patient in line with the COPE guidance.

Funding

None.

Conflict of Interest

None declared.

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