A 73-year-old woman presented with palpitations and right heart failure. Physical examination revealed elevated jugular venous pulse with a pansystolic murmur. ECG showed atrial flutter with a right bundle-branch block (Figure 1). A chest X-ray revealed normal cardiac size with a superior mediastinal opacity consistent with a goitre (Figure 2). Transthoracic and transoesophageal echocardiography demonstrated a large intramyocardial mass in the right ventricular freewall measuring up to 7.5×5 cm associated with right ventricular and right atrial dilatation, and systolic dysfunction (Figure 3A and B and online-only Data Supplement Movie I). The tricuspid valve was thickened with impaired leaflet mobility, resulting in severe tricuspid regurgitation (online-only Data Supplement Movie II). Tests to characterize the mass and to investigate other primary tumors were undertaken. A computed tomography scan confirmed a cardiac mass, a goitre, a small adrenal mass, and a renal cyst. Nuclear positron emission tomography scan revealed fluorodeoxyglucose uptake only within the heart (Figure 4). Cardiac magnetic resonance imaging (Figure 5A and online-only Data Supplement Movies III and IV) confirmed the solitary lesion and no other masses. Gadolinium contrast was not administered because of moderately severe renal dysfunction at the time of the cardiac magnetic resonance imaging. The mass was bright on T2 short tau inversion recovery sequence (Figure 5B). Urinary 5-hydroxyindoleacetic acid level was elevated. While waiting for definitive treatment, she succumbed to heart failure.

On autopsy, a well-circumscribed tumor, 8 cm in dimension, arising from the free wall of the right ventricle was detected (Figure 6). Histopathological examination revealed typical morphological and cytological features of a carcinoid tumor in the myocardium. There were islands of cells with intervening delicate-to-coarse fibrocollagenous stroma (Figure 7A). The tumor cells formed a conspicuous population of rosettes and pseudorosettes (Figure 7B). On immunohistochemistry, the tumor cells showed strong and diffuse positiv-
ity to neuroendocrine markers like chromogranin (Figure 7C). No other malignant primaries were identified on autopsy, specifically from the bowel and liver.

Carcinoid tumors are rare neuroendocrine malignancies most commonly arising from entero-chromaffin cells in the gastrointestinal tract. Carcinoid heart disease is often associated with carcinoid syndrome, which results in systemic symptoms, such as diarrhea and blushing, and is related to the excessive production of vasoactive substances such as 5-hydroxytryptamine. The most common features in cardiac carcinoid are tricuspid and pulmonary valve thickening and dysfunction, which leads to right-sided heart failure. The most common ECG finding is sinus tachycardia. Atrial flutter, as in our case, has not been reported. Furthermore, carcinoid heart disease presenting as a cardiac mass is rare, with only 4% in 1 series, and has previously been reported only with gastrointestinal and liver involvement. To the authors’ knowledge, isolated primary cardiac carcinoid heart disease without a gastrointestinal or liver primary, as highlighted in this case, has not been previously reported.

Disclosures

None.

References

Figure 4. Positron emission tomography with fluorine-18 fluorodeoxyglucose (FDG) tracer demonstrates increased but heterogeneous fluorine-18 fluorodeoxyglucose uptake within the heart (arrow). No additional sites of abnormal fluorine-18 fluorodeoxyglucose uptake are evident elsewhere in the body.

Figure 5. Cardiac magnetic resonance imaging in (A) apical 4-chamber view on balanced steady-state precession sequence demonstrating right ventricular mass and (B) T2 short tau inversion recovery sequence. The right ventricular mass (*) has a high signal.

Figure 6. A large mass (arrow) arising from the right ventricular free wall.
Figure 7. A. The tumor was arising from the myocardium (hematoxylin and eosin stain, ×10). B. Tumor cells formed rosettes and pseudorosettes (hematoxylin and eosin stain, ×40). C. Tumor cells stained strongly and diffusely for chromogranin on immunohistchemistry.
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