Utility of indium-111 octreotide to identify a cardiac metastasis of a carcinoid neoplasm

Mohammed Farooqui, MD, Sulaiman Rathore, MD, and Timothy Ball, MD, PhD

Carcinoid heart disease is classically described as right-sided valvular pathology. Solid cardiac metastases from carcinoid tumors are seldom reported. A multimodality imaging approach is needed to diagnose and localize this disease. Biopsy remains the gold standard to confirm the diagnosis of carcinoid. Octreotide uptake is characteristic of carcinoid tumor but not myxoma; thus, an indium-111 octreotide scan is very specific for the diagnosis of carcinoid tumor and helps in assessing the extent of carcinoid disease. We present a case in which an indium-111 octreotide scan revealed uptake in three distinct masses in the colon, liver, and right ventricle. The results of the scan were contradictory to the biopsy results, which were diagnostic for hepatic carcinoid and cardiac myxoma.

Figure 1. Multimodality imaging. (a) CT of the abdomen revealed a cardiac mass (☆) measuring approximately 5 cm involving the right ventricle and interventricular septum. (b) Transthoracic echocardiogram revealed a 5.2 × 6.2 cm homogeneous, circumscribed, infiltrating mass (☆) in the right ventricle. (c and d) Cardiac MRI demonstrated a smooth circumscribed mass (☆) in the right ventricular inflow tract, mid-cavity, and apex measuring 4.6 × 6.1 cm in the largest dimension, which enhances on T2 late gadolinium enhancement and signal loss with fat-saturated imaging.

Carcinoid tumors are rare and aggressive malignancies with a reported prevalence of 1.2 to 2.1 per 100,000 persons in the general population per year (1). Prompt recognition and diagnosis is of utmost importance (2). Solid carcinoid tumors rarely metastasize, and metastasis to the heart is even more rare (3). Carcinoid disease is diagnosed with a composite of clinical symptoms, 5-hydroxyindoleacetic acid assessment, appropriate imaging to localize the disease, and biopsy, which remains the confirmatory test. The telltale signs of carcinoid heart disease are involvement of right-sided heart valves with characteristic echocardiographic findings. Radionuclide scanning following intravenous ¹¹¹indium-labeled octreotide (¹¹¹In-DTPA-pentetreotide) provides a sensitive and noninvasive method of localizing somatostatin-positive tumors (somatostatin

receptor, SS-R expressed) and to monitor the efficacy of treatment (4–6).

An octreotide scan has an 86% sensitivity in detecting a carcinoid tumor (7). The false-positive rate for octreotide scans is around 12% and is mostly due to renal parapelvic cysts, accessory spleens, ventral hernias, or thyroid or breast disease (8). Many benign and malignant tumors are known to take up ¹¹¹indium-labeled octreotide, but uptake by myxomas has not been described in the literature. We report an intriguing case of a biopsy-confirmed myxoma with significant uptake on the indium-111 octreotide scan.

From Virginia-Tech Carilion School of Medicine and Research Institute, Roanoke, Virginia.

Corresponding author: Mohammed Farooqui, MD, 127 McClanahan Avenue, Suite 300, Roanoke, VA 24014 (e-mail: mafarooqui@carilionclinic.org).

Figure 2. Biopsy results. Liver biopsy with (a) synaptophysin immunohistochemistry and (b) hematoxylin and eosin staining consistent with neuroendocrine (carcinoid) cells. (c and d) Right ventricular tissue biopsy with hematoxylin and eosin staining depicting mucopolysaccharides suggestive of myxomatous tissue with no neuroendocrine cells, consistent with myxoma.

CASE REPORT

A 48-year-old woman presented to the emergency department with severe epigastric pain. Computed tomography (CT) of the abdomen revealed a liver mass and a cardiac mass involving the right ventricle (RV), measuring about 5 cm in diameter (Figure 1a). A transthoracic echocardiogram confirmed a 5.2 × 6.2 cm homogeneous, well-circumscribed, infiltrating RV mass (Figure 1b). The tricuspid valve was noted to be in close proximity to the tumor mass. The patient subsequently underwent cardiac magnetic resonance imaging (MRI), which showed evidence of a T2-enhancing RV mass (Figure 1c and 1d). Liver biopsy results were positive for carcinoid (Figures 2a and 2b).

Figure 3. A right coronary angiogram demonstrating hypervascularity and tumor perfusion from the right posterior descending and posterolateral coronary arteries.

Figure 4. An octreotide scan revealing indium-111 octreotide uptake by the right ventricular mass (black arrow) along with normal physiological uptake in the liver, spleen, kidneys, and urinary bladder.

Given the rarity of carcinoid metastasis to the heart, the possibility of two unrelated primary tumors was entertained, and an endomyocardial biopsy was performed to make the diagnosis. The patient underwent cardiac catheterization with coronary angiography (Figure 3), left and right heart catheterization, along with endomyocardial biopsy of the RV mass under fluoroscopic and intracardiac echocardiography guidance. The specimens obtained were adequate for analysis and revealed findings consistent with a benign cardiac myxoma (Figures 2c and 2d). To verify the diagnosis, an indium-111 octreotide scan was performed with the expectation that the liver mass would demonstrate uptake while the cardiac mass would

not. The octreotide scan revealed significant uptake by an undiagnosed mass in the cecum and in the liver mass; surprisingly, the RV mass also demonstrated significant octreotide uptake (Figure 4). This finding suggests a cecal primary with metastasis to both the liver and the RV. Treatment of the carcinoid tumor with octreotide was initiated, and the patient has been referred to determine if the cardiac tumor is suitable for resection.

DISCUSSION

This case demonstrates the ability of an indium-111 octreotide scan (9–11) to aid in defining the metabolic characteristics of multiple masses suspected of being carcinoid and to make a comprehensive assessment of the tumor burden, which is required for the development of a comprehensive treatment plan. More interestingly, this case reports a myxoma, confirmed histologically at endomyocardial biopsy, that demonstrated octreotide uptake on indium-111 octreotide scanning. Heretofore, no case of myxoma has been reported as demonstrating octreotide uptake. We believe this case represents a case of primary carcinoid tumor with metastasis to both liver and heart based on the results of octreotide scanning. Furthermore, this case widens the differential for RV mass lesions (12), suggesting the consideration of a metastatic carcinoid tumor that may present as a solid metastatic lesion to the heart.

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