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Authors: Aleksandra Niemiec-Pilch, Adam Maciejewski, Kosma Woliński, Paweł Gut, Piotr Stajgis, Marek Ruchała

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Isolated cardiac metastases of pulmonary carcinoid 13 years after resection of the primary tumor

Aleksandra Niemiec-Pilch¹, Adam Maciejewski¹, Kosma Woliński¹, Paweł Gut¹, Piotr Stajgis², Marek Ruchała¹

1. Department of Endocrinology, Metabolism and Internal Diseases, Poznan University of Medical Sciences, Poznan, Poland

2. Department of General Radiology and Neuroradiology, Poznan University of Medical Sciences, Poznan, Poland

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Correspondence:

Adam Maciejewski, M.D., Ph.D.
Department of Endocrinology, Metabolism and Internal Diseases
Poznan University of Medical Sciences, Poznan, Poland

Tel.: + 48 61 869 1330 Fax: + 48 61 869 1682

Mail: amaciejewski3@gmail.com

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Pulmonary neuroendocrine tumors (NETs) are a heterogeneous group of neoplasms. They can be divided into four categories: typical carcinoid, atypical carcinoid, and two high-grade subtypes - large cell neuroendocrine cancer and small cell lung carcinoma [1]. Pulmonary carcinoids (PCs) are rare, although the incidence has been observed to increase in recent decades. We describe a case of a woman with symptomatic isolated cardiac metastases of PC found 13 years after surgical resection of the primary tumor.

The patient was diagnosed with the right middle lobe lung tumor at the age of 44. In December 2004 she underwent lobectomy with lymphadenectomy. Histopathological examination revealed pulmonary carcinoid (T1N0M0, IA). She was then followed with annual chest radiography.

In May 2017, the patient was referred to the emergency department for syncope. She complained of progressive dyspnea and fatigue over the past few days. Moreover, hypotension was present on admission (the mean arterial pressure 50 mmHg). Computed tomography (CT) angiography (performed to exclude pulmonary embolism) revealed pericardial and pleural effusion with a pathologic mass of the interventricular septum (Figure 1A and 1B). To confirm the diagnosis of cardiac tamponade echocardiography was performed. It showed pericardial effusion (up to 20 mm), diastolic collapse of the right atrium and dilated inferior vena cava. The patient underwent pericardiocentesis to relieve symptoms. Cardiac magnetic resonance imaging (MRI) after recovery revealed two lesion - in the interventricular septum (Figure 1C) and another one in the basal anterolateral segments of the heart (Figure 1D). 18F-FDG PET/CT showed hypermetabolic areas corresponding to MRI findings any without other sites suggestive of malignancy. Pericardiotomy with biopsy proved typical PC cardiac metastases (Ki67 5-10 %).

The patient was qualified for somatostatin analog therapy after confirming somatostatin receptors overexpression in the cardiac metastases (Figure 1E and 1F). The
treatment started in September 2017. Since that time the patient has not presented any symptoms. Radiological control four months later demonstrated a response to the treatment (Figure 1G and 1H).

PCs may long be asymptomatic. Symptoms occurrence can result from local spread or metastases. An early manifestation is usually attributed to the secretion of humoral factors. Cardiac symptoms typically result from carcinoid heart disease [2]. The most common sites of PC metastasis are the liver, bone, adrenal gland, or brain. To date, about 40 cases of NET metastases to the heart have been described, most of them in small bowel NETs [3]. However, the frequency of NET cardiac metastases may be underestimated, as functional imaging studies revealed a frequency between 2.4 and 13 % [4,5]. Clinical presentation in such cases varies, from an asymptomatic course, through mild chronic symptoms (e.g., arrhythmias, exertional dyspnea), to severe life-threatening complications (acute heart failure, complete heart block, cardiac arrest, cardiogenic shock). Metastatic lesions are usually located in the ventricles or interventricular septum [3]. Cardiac metastases are usually expected in the case of multi-site metastatic disease.

This case points to a need for careful surveillance of NET patients, even many years after primary resection. Different imaging modalities, both morphologic and functional, may be needed.

References:


**Figure 1.** Imaging studies identifying pulmonary carcinoid metastases to the heart:

A - Computed tomography angiography of the pulmonary vessels showing pericardial effusion with thickness up to 16 mm around the right atrium (coronal plane); B – Computed tomography angiography of the pulmonary vessels: pathological mass (white arrow) situated in medial and apical segments of the interventricular septum (axial plane); C – Magnetic Resonance of the heart: pathological mass in medial and apical segments of the interventricular septum with maximum diameter of 31 mm (T2 TSE TRUFI sequence, short axis); D – Magnetic Resonance of the heart: pathological intrapericardial mass located in the basal segments of the anterolateral wall of the heart with maximum diameter of 37 mm (T2 TSE TRUFI sequence, short axis); E and F - 68Ga-DOTATATE positron emission tomography/computed tomography: mildly increased tracer uptake corresponding to lesions
found on cardiac magnetic resonance imaging (white arrows; axial plane); G and H – Magnetic Resonance of the heart (T2 TSE TRUFI sequence, short axis): examination after four months of therapy revealed a significant reduction in the size of pathological mass in the left ventricle (maximum diameter reduced to 22 mm) with a stable size of the second lesion.