Staged treatment of carcinoid syndrome complicated with severe tricuspid regurgitation. Author's reply

Marcin Waligóra[1, 2], Grzegorz Kopeć[1]

[1]Pulmonary Circulation Center, Department of Cardiac and Vascular Diseases, Faculty of Medicine, Jagiellonian University Medical College, John Paul II Hospital in Krakow, Poland

[2]Center for Innovative Medical Education, Department of Medical Education, Faculty of Medicine, Jagiellonian University Medical College, Kraków, Poland

Correspondence to:
Grzegorz Kopeć, MD, PhD, Pulmonary Circulation Center, Department of Cardiac and Vascular Diseases, Institute of Cardiology, Jagiellonian University Medical College, Prądnicka 80, 31–202 Kraków, Poland, phone: +48 12 614 33 99, e-mail: grzegorzkrakow1@gmail.com

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We thank Dr. Konsek-Komorowska and Prof. Pęczkowska for their interest [1] in our case report about a patient with carcinoid heart disease (CHD) [2]. We agree that management of CHD is complex, not always predictable, and requires multidisciplinary cooperation. While the largest cohort of patients with CHD including 240 operated patients presents a 33-year history of treatment approach [3], most current data come only from small case series or case reports [4]. Accordingly, it is a challenge to determine the preferred and optimal treatment approach in complex patients. In our patient, we decided to perform initially valve repair and then primary tumor extraction as the risk of abdominal surgery was considered too high.

At the time of diagnosis, the carcinoid tumor was in an advanced stage and the serum level of chromogranin A was 196 nmol/l (N: 0–6 nmol/l) and the urinary excretion of 5- hydroxyindoleacetic acid (5-HIAA) was 329.3 mg/24 h (N: 2–9 mg/24 h). Pathohistological grading was assessed as 2. Further examination of resected ileum showed proliferative activity of Ki67 (4%). Immunohistochemistry staining revealed a positive reaction to synaptophysin, chromogranin, and expression of somatostatin receptor 2 in all tumor cells. Staining for cytokeratin 7 and thyroid transcription factor 1 was negative. The overall staging was pT2 N1 PN1 LV1 M1c according to the American Joint Committee on Cancer (AJCC)/Union for International Cancer Control (UICC) 2017 classification (in order of appearance: primary tumor diameter of 2–5 cm, presence of regional lymph node metastasis, perineural invasion, invasion into lymphatic vessels, metastases to distant organs). The resected primary tumor appeared relatively small with a diameter of 3 cm without macroscopic infiltration to the ileal mucosa. There were no metastases to the appendix.

After abdominal surgery, N-terminal pro-B-type natriuretic peptide (NT-proBNP) level was slightly elevated to 182 pg/ml (<125). However, in the 12 months following surgery, the patient required reintroduction of low doses of loop diuretics due to progression of tricuspid regurgitation (which was observed first in September 2021: 6 months after cardiac and 2 months after abdominal surgeries).

A recent study has shown that 6.7% of patients operated for CHD needed cardiac re-intervention on follow-up due to tissue or mechanical valve dysfunction; however, only in 3 patients, a late valve dysfunction was due to carcinoid involvement of the artificial valve [3]. In our patient, we cannot definitively state the mechanism of biological valve dysfunction at least partly due to the lack of the results of 5-HIAA and chromogranin A, after primary tumor resection. However, we did not identify any other factors (i.e. infective endocarditis, pulmonary hypertension, right ventricle dilatation, etc.) that could contribute to bioprosthesis degeneration besides residual excretion of polypeptides by the tumor and its metastases [5]. Since the primary tumor resection, the tricuspid regurgitation, which was moderate at that point, did not progress in the following months. That supports a thesis that reduction of tissue mass prevented further valve damage. Currently, 18 months after cardiac surgery, the patient is treated with somatostatin analogs and telotristat ethyl. He is in New York Heart Association class II with

a NT-proBNP serum level of 983 pg/ml. Echocardiography shows stable moderate-grade tricuspid regurgitation and stable moderate dysfunction of the pulmonary valve (current mean gradient of 18 mm Hg vs. 15 mm Hg before cardiac surgery and PHT of 164 vs. 174 ms, respectively).

Article information

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