“Staged treatment of carcinoid syndrome complicated with severe tricuspid regurgitation” — Author’s commentary

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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, and 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 comes 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 tumour extraction as the risk of abdominal surgery was considered too high.

At the time of diagnosis the carcinoid tumour was in advanced stage and a 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 positive reaction to synaptophysin, chromogranin and expression of somatostatin receptor 2 in all tumour cells. Staining for
cytokeratin 7 and thyroid transcription factor 1 were negative. The overall staging was pT2 N1 PN1 LV1 M1c according to American Joint Committee on Cancer (AJCC)/Union for International Cancer Control (UICC) 2017 classification (in order of appearance: primary tumour diameter of 2–5 cm, presence of: regional lymph node metastasis, perineural invasion, invasion into lymphatic vessels, metastases to distant organs). The resected primary tumour appeared as 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 following 12 months after 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 two months after abdominal surgeries).

A recent study has shown that 6.7% of patients operated for CHD needed cardiac re-intervention in 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 make a certain statement on the mechanism of biological valve dysfunction, it is at least partly due to the lack of the results of 5-HIAA and chromogranin A, after primary tumour 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 tumour and its metastases [5]. Since the primary tumour resection, the tricuspid regurgitation which was moderate at that point, did not progress in next months. That advocate for thesis that reduction of tissue mass prevented further valve damage. Currently, 18 months after cardiac surgery the patient is treated with somatostatin analogues and telotristat ethyl. He is in New York Heart Association class II with the 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).

References


