Rare cardiac tumors represent an ultimate challenge for the whole Heart Team. Authors' reply

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DOI: 10.33963/KP.a2022.0038

Received:
January 28, 2022

Accepted:
February 4, 2022

Early publication date:
February 7, 2022

We are very delighted that our recently published article in which we presented a peculiar case of a primary cardiac neuroendocrine tumor [1] drew the attention of the scientific community and wish to thank readers for their interest in our work. This uncommon disease, which has been encountered at our center for the first time, initially left our whole team confused. Nevertheless, the detailed multimodal imaging and precise diagnostics cleared the way for safe and successful surgical removal, which together with comprehensive postoperative pathohistology, immunohistochemistry analyses, and additional imaging workup, enabled the patient's appropriate overall management.

In their comment, Peczkowska et al. [2] provide a very practical and methodical overview of paragangliomas. It includes their pathogenesis and classification, with special emphasis on the potential genetic basis of the disease mainly due to the succinate dehydrogenase (SDH) mutation variants that can be identified in three-quarters of patients with cardiac paragangliomas [3]. The authors [2] highlighted the importance of genetic testing, particularly for SDH genes, for determining the hereditary background of the disease in such patients. Our local healthcare system currently does not have the resources that could provide the aforementioned genetic testing, so unfortunately it could not have been performed in our patient [1]. Based on the information taken at the time of admission, as well as detailed family history investigated subsequently after the final diagnosis, we could confirm that the patient's family history was negative. Nonetheless, we do acknowledge that the negative family history cannot exclude the hereditary background of the disease due to the variable penetrance of the mutations [4].

Regarding the metastatic potential of the tumor, as part of the systemic evaluation, our patient underwent comprehensive and detailed imaging including octreotide scan, computed tomography of the chest, and magnetic resonance imaging of the abdomen and small pelvis, which were all negative for other tumor foci. Some additional analyses that might have been considered include metaiodobenzylguanidine (MIBG) scan and hormonal examinations. However, these were not performed because the patient did not present any symptoms whatsoever that would be suggestive of excessive catecholamine production (i.e. hypertension, headaches, pallor, excessive sweating), nor any other abnormal hormonal activity such as carcinoid syndrome. To the present day, more than two years later and with regular periodic follow-up, there have been no signs of distant metastases or local disease recurrence in our patient.

We would like to emphasize that this challenging cardiac tumor could not have been managed properly without the remarkable cooperation and teamwork of a wide array of different medical specialties. The Heart Team, consisting of a cardiologist and a cardiac surgeon together with an anesthesiologist, was crucial for establishing the right approach and making the decision for surgical removal of the tumor. The surgery itself was performed in general endotracheal anesthesia through

total median sternotomy. Pericardiotomy revealed normal heart in sinus rhythm, situs solitus, with a large, pale pink tumor that was present on the lateral side of the right atrium and ventricle. Further surgery was performed on total cardiopulmonary bypass (total heparinization, selective bicaval to the ascending aorta, venting through the ascending aorta), using intermittent antegrade hyperkalemic solution (St. Thomas). The tumor sample was sent for *ex tempore* and imprint cytology analyses when the diagnosis of the neuroendocrine tumor was suspected. The tumor mass was then excided *in toto* from the heart walls and the right coronary artery around which it was wrapped. Total aortic cross-clamping time was 58 minutes, with a total bypass time of 80 minutes. Following the surgery, it was up to the pathologist, radiologist, nuclear medicine specialist, and oncologist to establish the final diagnosis and perform additional examinations and patient follow-up.

In essence, cardiac involvement is not unusual in patients with carcinoid syndrome due to systemic effects of neuroendocrine tumors (characteristically represented by carcinoid heart disease — a fibrous thickening of the endocardium, mainly involving the heart valves, leading to their malfunction) [5]. However, the primary localization of neuroendocrine tumors in the heart is extremely rare and requires the outstanding cooperation of different medical specialists for proper patient management and favorable outcome.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

Conflict of interest: None declared.

REFERENCES

1. Redzek A, Preveden A, Todic M, et al. Primary neuroendocrine tumor of the heart. Successful management of an extremely rare disease. Kardiol Pol. 2022; 80(2): 226–228, doi: 10.33963/KP.a2021.0188, indexed in Pubmed: 34936083.
2. Peczkowska M, Konsek-Komorowska S. Cardiac paraganglioma — a challenging diagnostic and treatment dilemma. Kardiol Pol. 2022; 80(3): 378–379, doi: 10.33963/KP.a2022.0022, indexed in Pubmed: 35114002.
3. Martucci VL, Emaminia A, Del Rivero J, et al. Succinate dehydrogenase gene mutations in cardiac paragangliomas. Am J Cardiol. 2015; 115: 1753–1759.
4. Hoekstra AS, Devilee P, Bayley JP. Models of parent-of-origin tumorigenesis in hereditary paraganglioma. Semin Cell Dev Biol. 2015; 43: 117–124.
5. Davar J, Connolly HM, Caplin ME, et al. Diagnosing and managing carcinoid heart disease in patients with neuroendocrine tumors: an expert statement. J Am Coll Cardiol. 2017; 69(10): 1288-1304.