Primary neuroendocrine tumor of the heart. Successful management of an extremely rare disease

Aleksandar Redzek[1, 2*], Andrej Preveden[1, 2*], Mirko Todic[1, 2], Nikola Komazec[2], Milena Spirovski[1], Golub Samardzija[1, 2], Mihaela Preveden[1, 2], Ranko Zdravkovic[2], Vanja Drljevic Todic[1, 2], Anastazija Stojsic Milosavljevic[1, 2]

[1]Faculty of Medicine, University of Novi Sad, Novi Sad, Serbia
[2]Institute of Cardiovascular Diseases Vojvodina, Sremska Kamenica, Serbia
[*]Both authors equally contributed to the study

Correspondence to:
Andrej Preveden, MD, Institute of Cardiovascular Diseases Vojvodina, Put dr Goldmana 4, 21204 Sremska Kamenica, Serbia, phone: +38 121 480 57 02, e-mail: andrej.preveden@mf.uns.ac.rs

DOI: 10.33963/KP.a2021.0188

Received:
October 26, 2021

Accepted:
December 21, 2021

Early publication date:
December 22, 2021

A 38-year-old female without medical history was admitted due to severe chest pain. She had no prior symptoms, and this was the first presentation of the disease. Blood pressure was normal. Electrocardiography showed ST depression in inferolateral leads (Figure 1A). High-sensitive troponin I was elevated — 1571.5 ng/l (normal <50 ng/l).

Echocardiography detected a tumor near the right atrium (Figure 1B; Supplementary material, *Video S1*). Additional imaging with computed tomography and magnetic resonance (Figure 1C–E) showed that the tumor was hypervascular, non-invasive, and well-contained mass. It was in close contact and compressing the right atrium and right ventricle but without infiltrating them. It surrounded the right coronary artery. A coronary angiogram revealed no lesions, so coronary artery disease was excluded. Based on radiological features, the neuroendocrine tumor was primarily suspected with a possible differential diagnosis indicating paraganglioma.

Surgical removal was recommended by the heart team. Surgery was performed through median sternotomy using cardiopulmonary bypass (Figure 1F). The tumor was excised *in toto* from the heart walls and the right coronary artery.

The tumor was 55 × 39 × 35 mm in size (Supplementary material, *Figure S1A*). Microscopically, the tissue was composed of uniform polygonal cells with round nuclei, chromatin was fine-grained and diffusely distributed, and cytoplasm was pale eosinophilic. Tumor cells were arranged in organoid, trabecular, and pseudo-glandular formations (Supplementary material, *Figure S1B*). The stroma of the tumor was poor, built of thin connective tissue bands with numerous small blood vessels. Tumor necrosis was absent, and mitoses were rare (<2/10 HPF). The Ki67 proliferative index was <1% (Supplementary material, *Figure S1C*). The immunohistochemical profile was positive for CD56, synaptophysin, and chromogranin (Supplementary material, *Figure S1D–F*). Based on these analyses, the diagnosis of a typical neuroendocrine tumor was made.

The postoperative course was uneventful, and the patient was discharged on the 8th postoperative day. Subsequently, an octreotide scan, computed tomography of the chest, and magnetic resonance of the abdomen and small pelvis were performed in search of other potential tumor localizations. All these studies were negative, so this was a primary tumor of the heart. To the present day, two years after the initial presentation, the patient has been free of symptoms and with no signs of disease recurrence.

Solitary neuroendocrine tumors are rarely found in heart structures. The majority of these are metastatic tumors of the gastrointestinal origin, particularly the small intestine [1]. However, the primary localization of neuroendocrine tumors in the heart is extremely rare [2, 3]. Our case is unique because the tumor was located on the lateral wall of the right heart surrounding the right coronary artery, which contributed to the complexity of surgical management.

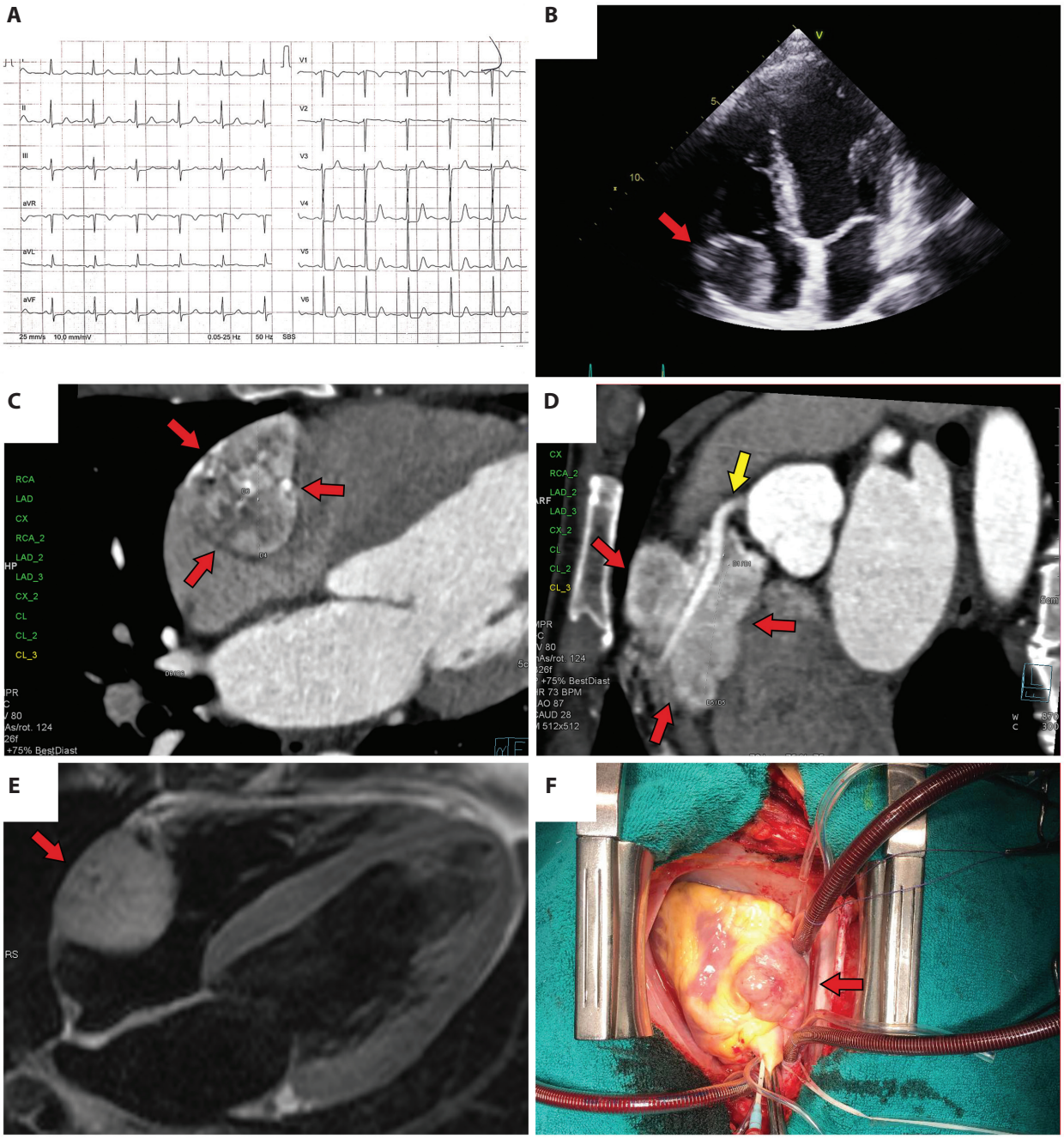

Figure 1. A. Electrocardiography on admission: ST depression in leads II, III, aVF, V4–V6. **B.** Transthoracic echocardiography, apical 4-chamber view: the tumor (red arrow) is located near the right atrium and collapsing it. **C–D.** Cardiac computed tomography: the tumor (the red arrows) is compressing the right atrium and the right ventricle but not infiltrating them and tightly surrounds the right coronary artery (yellow arrow). **E.** Cardiac magnetic resonance: turbo spin-echo (T2)-weighted image shows a hyperintense soft tissue lesion (red arrow). **F.** Intraoperative view: tumor (red arrow) is present on the lateral side of the right atrium and ventricle

The final diagnosis in our patient was a neuroendocrine tumor. Most likely differential diagnosis includes paraganglioma, which is regarded as a sub-family of neuroendocrine neoplasms [4]. This is indicated by the tumor's location, radiological features, the absence of metastatic disease, as well as histology and immunohistochemistry findings. However, the clinical presentation did not suggest any hormonal activity of the tumor, and the patient had a negative family history, while paragangliomas can be hereditary in up to 50% of cases [5].

Supplementary material

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

Article information

Conflict of interest: None declared.

REFERENCES

1. Jann H, Wertenbruch T, Pape U, et al. A matter of the heart: myocardial metastases in neuroendocrine tumors. Horm Metab Res. 2010; 42(13): 967–976, doi: 10.1055/s-0030-1267204, indexed in Pubmed: 20972943.
2. Kavesh MH, Drew PA, Stewart BD. Primary cardiac low-grade neuroendocrine tumor: a case report and review of the literature. Cardiovasc Pathol. 2021; 52: 107317, doi: 10.1016/j.carpath.2021.107317, indexed in Pubmed: 33434636.
3. Altshuler E, Saker H, Ramnaraign B. Primary neuroendocrine tumours of the heart: case report and literature review. BMJ Case Rep. 2021; 14(6), doi: 10.1136/bcr-2021-242517, indexed in Pubmed: 34116995.
4. Rindi G, Klimstra DS, Abedi-Ardekani B, et al. A common classification framework for neuroendocrine neoplasms: an International Agency for Research on Cancer (IARC) and World Health Organization (WHO) expert consensus proposal. Mod Pathol. 2018; 31(12): 1770–1786, doi: 10.1038/s41379-018-0110-y, indexed in Pubmed: 30140036.
5. Michałowska AM, Ćwikła JB, Konka M, et al. Eleven-year follow-up of cardiac paraganglioma in a patient with SDHD C11X gene mutation. Kardiol Pol. 2021; 79(11): 1276–1277, doi: 10.33963/KP.a2021.0085, indexed in Pubmed: 34392514.