POLISH HEART JOURNAL

Kardiologia Polska
The Official Peer-reviewed Journal of the Polish Cardiac Society since 1957

Online first
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ISSN 0022–9032 e-ISSN 1897–4279

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Article type: Clinical vignette

Received: January 27, 2025

Accepted: February 11, 2025

Early publication date: February 28, 2025

Primary leiomyosarcoma of the right ventricle with metastases in a young female

Short title: Primary leiomyosarcoma of the right ventricle

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Primary heart tumors (PHT) are an infrequent phenomenon affecting 0.02% of the population; among them, 25% are malignant tumors [1]. The most common PHT is angiosarcoma, while leiomyosarcoma accounts for less than 1% [2].

A 48-year-old female patient with a history of lower limb venous thrombosis and hysterectomy due to benign smooth muscle tumors was transferred to our department due to a tumor in the right ventricle found on transthoracic echocardiography (Figure 1A) and suspicion

of metastases to the lungs, spine, and kidneys (Figure 1B). Nine days earlier, the patient was admitted to the emergency room with progressively worsening dyspnea, significantly increased fatigue, and exertional intolerance. On admission, she was classified as III/IV class in a New York Heart Association scale without angina. Physical examination revealed lower limb edema, a heart murmur, and basilar lung rales. The patient underwent additional cardiac magnetic resonance (CMR) assessment, showing a tumor in the right ventricle (Figure 1C), and was consulted with oncologists.

Finally, after the Heart Team discussion, she was qualified for tumor resection with the collection of material for histopathological examination as a palliative life-saving procedure.

The patient was successfully operated on under general anesthesia. The tumor was removed in two fragments (Figure 1D–F). A biological valve implant (C-E Perimount 31 mm) was also needed in the tricuspid position. Postoperatively, a dual-chamber pacemaker was implanted due to a complete atrioventricular block.

Histopathological examination revealed leiomyosarcoma without signs of necrosis, classified by French Fédération Nationale des Centres de Lutte Contre (FNCLCC) as Grade 2, without evidence of angioinvasion (LVI0) and neuroinvasion (Pn0).

After hospital discharge and rehabilitation, the patient received systemic chemotherapy according to ADIS protocol (dacarbazine + doxorubicin).

Leiomyosarcoma may remain asymptomatic while consistently growing. At the time of diagnosis, it is often infiltrating the heart wall and is linked with metastases [4]. While malignant tumors present 14.1 months of estimated median survival time in case of localization in the right heart or stage 4, leiomyosarcoma decreases it to only two months in patients with other organs involved [3, 5].

Interestingly, according to previous reports, there were patients who before symptoms of leiomyosarcoma of the heart, had been diagnosed with uterine leiomyoma or had uterine fibroids removed, like our patient [5].

There is, therefore, a possibility that the tumor in our patient's heart was not a primary lesion.

This assumption is consistent with the involvement of the inferior vena cava, which is often the first place this generator produces output but most often metastasizes through continuity to the right atrium.

Another possibility is the existence of a rare genetic predisposition connecting these dots. Further research might shed more light on this topic.

Article information

Conflict of interest: None declared.

Funding: None.

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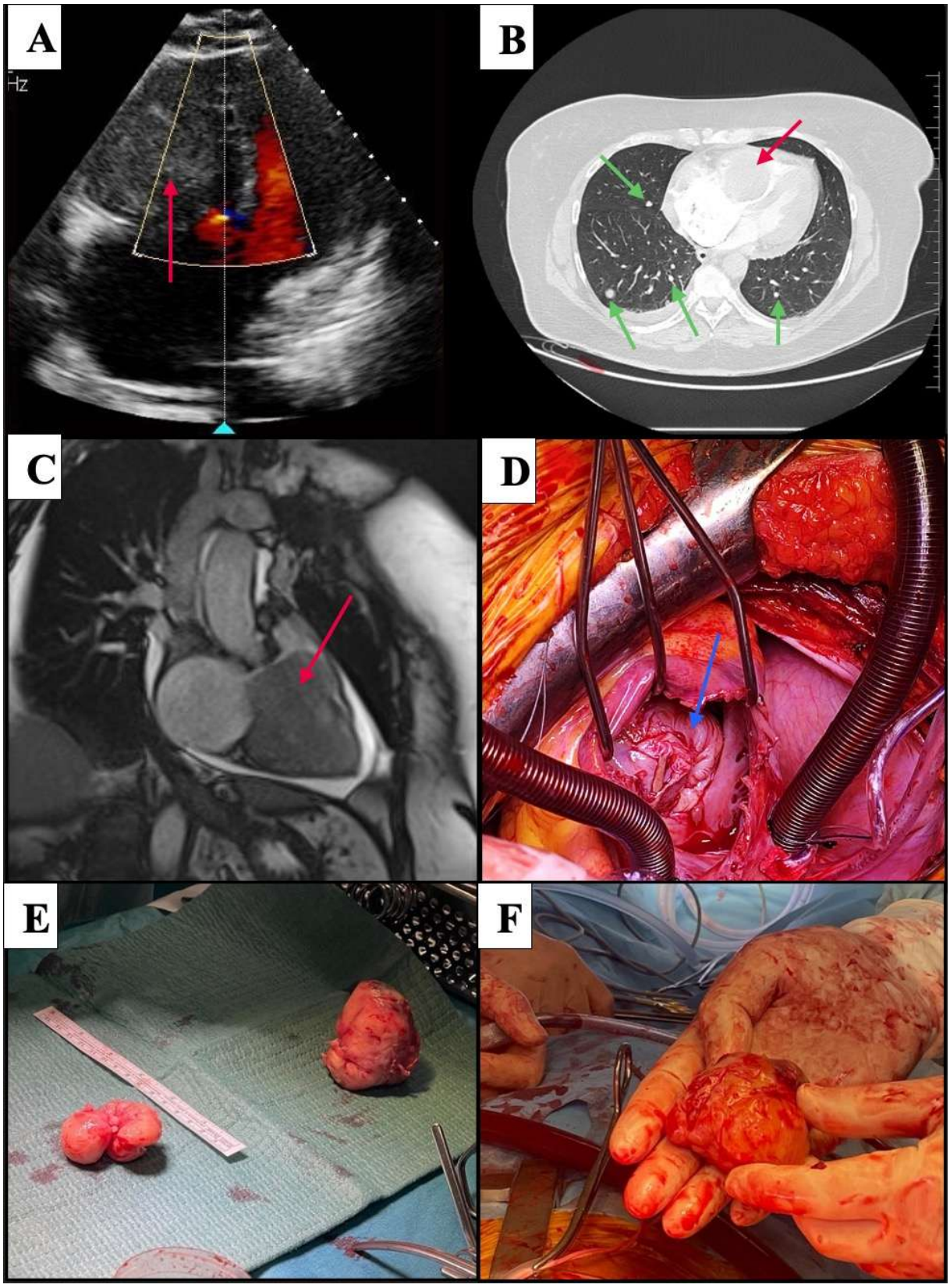

Figure 1. A. Echocardiography showing enlarged right ventricle with flow obturation due to enormous tumor (red arrow). **B.** Computed tomography scan of the tumor (red arrow) also showing multiple metastases (green arrows) and fluid retention in the lungs. **C.** Magnetic resonance imaging shows tumor mass (red arrow). **D.** Intraoperative photo of the tumor placed

in the right ventricle (blue arrow). **E.** Photo of two pieces of the tumor. **F.** Intraoperative photo of the first part of the tumor