A 72-year-old Japanese woman was admitted to hospital because of worsening general condition, dyspnea, and flank pain. She had a history of treatment for gastrointestinal stromal tumor (GIST), involving partial gastrectomy 10 years earlier. Laboratory analysis indicated hematuria and high serum lactate dehydrogenase (1,806 units/L). Chest X-ray did not indicate pulmonary artery dilatation. Electrocardiogram showed complete right bundle branch block, left atrial overload, and negative T wave in the leads (Figure S1). Echocardiography showed an immobile mass in the cavity in front of the interventricular septum (arrowheads). (B) Four-chamber cardiac magnetic resonance imaging, showing a huge mass with hypointense signal occupying the whole right ventricle and extending to the right atrium (RA; arrowheads). (C-E) Histopathology of the resected tumor indicated (C) proliferation of spindle cells (HE), which were positive for both (D) CD34 and (E) KIT immunostain, suggesting that the mass was metastatic gastrointestinal stromal tumor. Ao, aorta; LA, left atrium; LV, left ventricle.
imaging showed a huge mass with hypointense signal occupying the whole RV and extending to the right atrium (RA; Figure 1B).

Although the cardiac biopsy was considered at first for definitive diagnosis, it was not performed because of high risk of sudden death and pulmonary embolism. Under these conditions, biopsy from the rib, where computed tomography (CT) showed the presence of tumor, confirmed the GIST histology. Taking into account the clinical course and the histological findings of rib tumor, the huge mass in the RV was strongly suspected to be metastatic GIST.

If the huge tumor in the RV was metastatic GIST, molecular target drugs would be expected to have a high therapeutic effect. More than 90% of Japanese patients with unresectable and/or metastatic GIST undergoing imatinib mesylate therapy have had partial response or stable disease. The median progression-free survival time is 96 weeks and overall survival estimates are 73.6% at 3 years.

Imatinib mesylate therapy was therefore planned after the surgical debulking of the RV mass, which was done to confirm the histological diagnosis as well as prevent obliteration of the cardiac chamber and valve obstruction. Histopathology of the resected tumor indicated proliferation of spindle cells (Figure 1C) which were positive for both CD34 and KIT immunostain (Figure 1D,E), suggesting that the mass was metastatic GIST. GIST is one of the most common sarcomas of the gastrointestinal tract, and rarely metastasizes to the heart. There have been no reports of metastatic GIST in the RV.

Pulmonary hypertension was not present in the right heart catheter after surgery. Moreover, contrast-enhanced CT as well as lung ventilation-perfusion scintigraphy did not show pulmonary embolism (Figure S2).

The condition caused by the occupation of the RV by metastatic GIST was successfully improved by the surgical procedure, which was associated with a reduction in systolic RV pressure estimated on echocardiography from >50 down to 21.0 mmHg, although a large mass still remained in the RV (Figure 2A,B). Two months later, imatinib mesylate therapy was started. The patient had partial response 3 months after imatinib therapy. Contrast-enhanced CT showed substantial volume reduction of tumor in the RV (Figure 2C,D).

We present here imaging of an unusual huge metastatic GIST occupying the RV. Definitive diagnosis, including histopathology, is critical for the treatment of cardiac tumor.

Disclosures
The authors declare no conflicts of interest.

References

Supplementary Files

Supplementary File 1

Figure S1. Electrocardiogram showing complete right bundle branch block, left atrial overload, and negative T wave in the leads.

Figure S2. Lung ventilation-perfusion scintigraphy showing absence of pulmonary embolism.

Please find supplementary file(s); http://dx.doi.org/10.1253/circj.CJ-17-0722