A case of dedifferentiated liposarcoma of the heart and stomach

Yoichi Hisata a,⁎, Yuichi Tasaki a, Satoshi Kozaki b, Takafulmi Yamada a

a Division of Cardiovascular Surgery, Oita Prefectural Hospital, 476 Bunya, Oita City, Oita 870-8511, Japan
b Division of Cardiovascular Surgery, Almeida Memorial Hospital, 1509-2 Miyazaki, Oita City, Oita 870-8133, Japan

ARTICLE INFO

Article history:
Received 9 July 2017
Received in revised form 3 October 2017
Accepted 4 October 2017
Available online 6 October 2017

Keywords:
Liposarcoma
Dedifferentiated type
Cardiac tumor

ABSTRACT

INTRODUCTION: Liposarcoma of the heart and stomach is rare.
PRESENTATION OF CASE: We report a case of liposarcoma in both organs with dedifferentiated histology. A patient was referred to our hospital with anorexia and weight loss. Upper gastrointestinal tract endoscopy revealed 5–10 mm elevated lesions, and echocardiography and computed tomography showed tumorous lesions in the left atrium. Tumor resection and mitral valve replacement were performed, and biopsy was performed for the gastric tumor. Both the tumors were diagnosed as dedifferentiated liposarcoma.
DISCUSSION: Liposarcoma – a mesenchymal malignant tumor that contains lipoblasts – is the second most common soft tissue sarcoma. The tumor occurs most frequently in the limbs and retroperitoneum and rarely originates in the heart and the stomach.
CONCLUSION: Chemotherapy and radiotherapy are only adjunctive therapies but not as standard treatment for cardiac tumors. Therefore, we believe that wide surgical resection was the best choice of treatment in the present case.

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1. Introduction

Liposarcoma is the second most common malignant soft tissue tumor. The tumor occurs most frequently in the limbs and retroperitoneum, and rarely originates in the heart and stomach. Histologically dedifferentiated liposarcoma is an exceedingly rare neoplasm in the heart. Herein, we report a rare case of dedifferentiated liposarcoma of the heart and stomach. This paper has been written in line with the SCARE criteria [1].

2. Case report

A 79-year-old woman presented with anorexia and weight loss. Upper gastrointestinal tract endoscopy revealed 5–10 mm elevated lesions in the greater curvature of the gastric angle. Echocardiography and computed tomography (CT) showed tumorous lesions in the left atrium; the lesions connected to left atrial roof, atrial septum, and left atrial floor from the mitral valve anterior leaflet nearly incarceration. The lesion in the tumors showed low or mixed echogenicity. At first, these findings suggested that malignant lymphoma was the most likely diagnosis.

Moreover, there was mitral valve stenosis, with an area of 1.3 cm². No other tumorous lesions were detected on head, chest, and abdominal CT (Fig. 1).

The patient underwent surgery for the cardiac tumor. A superior septal approach was used to access the left atrium. The tumor continued from the mitral valve anterior cusp to the left atrial roof and atrial septum. We used blunt dissection and removed the tumors. We resected them as much as possible, but were unable to remove them completely.

Resection of the tumors, adhering to the mitral valve anterior leaflet led to a loss of mitral valve coaptation; therefore, we performed mitral valve replacement using a 23-mm Carpentier-Edwards Perimount (Edwards Lifesciences, Irvine, Calif). Macroscopically, the tumor was a 5-cm elastic, hard mass with an uneven white surface. Histopathological examination showed infiltrative growth of spindle-shaped atypical cells, sporadically associated with mitosis and multinucleated cells. Hemangiopericytomaticus vessels were partially observed. Immunostaining showed positivity for cdk4, MDM2, and S-100. According to the WHO classification, the tumor was a dedifferentiated liposarcoma (Fig. 2). Histopathological examination failed to identify the primary lesion.

The patient was discharged from the ICU 8 days postoperatively. Unfortunately, she died of septic shock due to urinary tract infection 55 days postoperatively.

3. Discussion

Liposarcoma – a mesenchymal malignant tumor that contains lipoblasts – is the second most common soft tissue sarcoma. The tumor occurs most frequently in the limbs and retroperitoneum and rarely originates in the heart and the stomach.
Fig. 1. **Top left**, Upper gastrointestinal tract endoscopy revealed multiple protruding lesions (*). **Bottom left**, CT demonstrating multiple tumors (**) in the left atrium. **Top right**, Echocardiogram indicating mitral valve stenosis and a tumor (***) (**). **Bottom right**, Echocardiogram showing that the tumor (****) extended to the vicinity of the aortic root.

Fig. 2. **Top left**, An intraoperative photograph showing a tumor (*) with an irregular surface that extended to the mitral valve anterior leaflet. **Bottom left**, resected tumor specimen; the mitral valve anterior leaflet (**) is located at the right end. **Pathological photomicrograph of the resected specimens from the stomach (top middle) and heart (bottom middle) showing infiltrative growth of spindle-shaped cells with marked nuclear atypia (hematoxylin & eosin staining).** The neoplastic cells showing positivity for MDM2 (top right) and CDK4 (bottom right) (immunostaining).
Neragi-Miandoab et al. reported only one case with liposarcoma among 117 patients with malignant cardiac tumors [2]. He et al. histopathologically reviewed the English literature and found only one case had dedifferentiated liposarcoma among 31 cases [3].

The rate of cardiac metastasis of soft-tissue sarcoma is 8–25%. Myocardial metastasis accounts for 50% of cardiac sarcoma metastasis cases, and hematogenous metastasis is common [4]. Furthermore, the gastric liposarcoma was rare at only 9.6% of 6998 patients with soft tissue sarcoma [5].

Based on these findings, cardiac liposarcoma described herein is speculated to be a hematogenous metastasis from gastric liposarcoma, though this is difficult to prove with certainty. Similar to other cardiac malignant tumors, liposarcoma has a poor prognosis, with survival of 2 months to 2 years. For gastric liposarcoma, surgical resection is the first choice, and total or subtotal gastrectomy is usually performed. However, the prognosis is poor because of a high recurrence rate. The patient reported herein died of septic shock 2 months postoperatively. Chemotherapy and radiotherapy are only adjunctive therapies but not established as standard treatment for cardiac tumors. However, tumor resection allows: 1) mechanical reduction of tumor size, 2) histopathological diagnosis, and 3) mitral valve stenosis treatment and incarceration prevention. Therefore, we believe that wide surgical resection was the best choice of treatment in the present case.

Conflicts of interest
None.

Funding
None.

Ethical approval
Not applicable.

Consent
None declared.

Author contribution
Study concept; Y. Hisata, Y. Tasaki, S. Kozaki and T. Yamada.
Data collection; Y.Hisata.
Writing the paper; Y.Hisata.

Guarantor
Takafumi Yamada, the corresponding author of this paper.

Acknowledgment
The authors would like to thank Shogo Urabe, MD, of the Department of Pathology, Oita Prefectural Hospital, for pathological assistance.

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