Case report - Cardiac general
Large primary cardiac sarcoma on the left ventricular free wall: is total excision contraindicated?

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Abstract

A case of a large primary cardiac sarcoma on the left ventricular free wall is reported. Although the definitive diagnosis of this tumor was not made preoperatively, total excision was planned for rapid diagnosis and optimal procedure. However, the operation was discontinued due to intraoperative diagnosis of malignancy. As a result, the patient suffered from the symptoms of cardiac tamponade caused by the large tumor. We discuss the surgical strategy to provide therapeutic benefit for possible patients in the future. In conclusion, an aggressive attempt at volume reduction such as cardiac autotransplantation may relieve the symptoms, even though such surgery would only be palliative.

Keywords: Cardiac tumors; Sarcoma

1. Introduction

Primary cardiac tumors are uncommon entities with an extremely low incidence [1]. The majority of primary cardiac tumors are benign atrial myxomas that can be surgically excluded. Similarly, surgical excision is the most effective local treatment for primary cardiac malignancies [2]. However, surgical outcome was very poor in spite of additional treatments, such as radiotherapy and chemotherapy [3]. In 2006, Reardon et al. reported a new challenging procedure (cardiac autotransplantation; cardiac explantation and ex vivo tumor resection with cardiac reconstruction) for complete excision of complex left-sided tumors [4]. However, the surgical strategy for malignant cardiac tumors has not been standardized due to individual variations of the tumors in size, location, and origin. Thus, it is very important to consider the optimal surgical strategy for each patient.

A case of a large primary cardiac sarcoma on the left ventricular (LV) free wall is reported. Although the definitive diagnosis of this tumor was not made preoperatively, total excision was planned for rapid diagnosis and optimal procedure. However, the operation was discontinued due to intraoperative diagnosis of malignancy. As a result, the patient suffered from the symptoms of cardiac tamponade caused by the large tumor. We discuss the surgical strategy for primary cardiac malignancies to provide therapeutic benefit for possible patients in the future.

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2. Clinical summary

A 73-year-old man was referred to our hospital due to general fatigue and severe weight loss (14 kg over three months). Chest computed tomography showed a large avascular tumor (10 × 14 × 16 cm³) on the left side of the heart (Fig. 1a). The content of the tumor was relatively homogenous, but included some septum. The borderline between the tumor and the LV was unclear. Two-dimensional echocardiography revealed that the tumor, which included multiple cystic lesions, severely compressed the heart and caused cardiac tamponade (Fig. 1b). Coronary angiography showed many small arteries going towards the tumor from the diagonal branch. There were no lesions that were suspected of distant metastases in other organs. Thus, total excision of the large tumor was planned for rapid diagnosis and optimal treatment.

Under general anesthesia, the chest was entered via a median sternotomy. The pericardium was severely adherent to the heart and the tumor. During dissection of the tumor, a large amount of serous effusion appeared when the tumor was injured. Necrotic tissue was found in the tumor (Fig. 2a). The tumor was resected for rapid frozen-section diagnosis. The dysplastic cells of the specimen suggested malignancy. Although the origin of the tumor was not detected, the operation was discontinued due to severe adhesions surrounding the malignant tumor from which there was no foreseeable cure.

Although the patient was extubated on the first postoperative day, the patient’s respiratory condition deteriorated progressively over a week. Despite postoperative
chemotherapy with ifosfamide, the patient died on the 31st day. The definitive pathological diagnosis was undifferentiated, high-grade, pleomorphic sarcoma (Fig. 2b). The postmortem macroscopic specimen suggested that the origin of the sarcoma was the LV epicardium (Fig. 2c).

3. Discussion

Due to the rarity of primary malignant cardiac tumors, surgical strategies (i.e., indications, methods, and approaches) have not been well established. Had the tumor been diagnosed preoperatively as malignant in this case, many surgeons would have considered that surgery was not indicated. In fact, however, the diagnosis was not made preoperatively. A definitive diagnosis by needle or direct biopsy would have been required prior to total excision, but such a biopsy is associated with higher risks for surgical complications, such as uncontrollable hemorrhage, cardiac tamponade, and dissemination. To relieve the severe symptoms of cardiac tamponade caused by the undiagnosed tumor, total excision of the tumor for both rapid diagnosis and optimal treatment was planned.

For such patients with a malignant cardiac tumor, short-term quality of life is one of the highest priorities due to the poor long-term prognosis. Although total excision would be the optimal treatment, any operation should result in relief from symptoms following surgery. Thus, tumor volume reduction would be an alternative to alleviate severe symptoms. However, we discontinued the operation because a cure was not possible. Furthermore, the large intrapericardial space occupied by the tumor was one of the greatest limitations for further surgical maneuvers. With an extremely large tumor that had grown up the LV free wall, in vivo total excision would be unfeasible. However, a better surgical procedure should have been selected once the operation had started.

An earlier surgical report demonstrated that a technique of cardiac autotransplantation (cardiac explantation, ex vivo tumor resection with cardiac reconstruction) is a challenging method for treating primary cardiac tumors [4]. In retrospect, this technique would have been appropriate, because the LV epicardium may have been the origin of the tumor. As shown in Fig. 2c, ex vivo volume reduction of the tumor would have been feasible without injury to the LV myocardium. Although this technique for volume reduction of a primary malignant cardiac tumor has an extremely high-risk, including surgical hemorrhage from the resection, the short-term quality of life would have been much better than with the surgical treatment provided.

4. Conclusion

Better short-term quality of life is the highest priority for patients with primary cardiac malignancy, which is associ-
ated with a poor long-term prognosis. Accordingly, an aggressive attempt at volume reduction has a chance to relieve the symptoms caused by a malignant cardiac tumor, even though such surgery would only be palliative.

References


