Radical surgery for a mitral valve intimal sarcoma: serial patch repair of the annulus and atrium

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Abstract

We report the rare case of a 48-year-old woman with a cardiac intimal sarcoma occupying the mitral valve and the left atrial roof. We resected the tumour in one block without residual lesions. The anterior mitral annulus and left atrial roof were serially repaired with the bovine pericardium, and a mechanical prosthesis was implanted in the reconstructed mitral annulus. The patient remains alive without recurrence for more than 1 and a half years postoperatively. We performed radical surgery for a mitral valve intimal sarcoma, and serial patch repair of the anterior mitral annulus and left atrial roof was a useful technique.

Keywords: Cardiac intimal sarcoma • Radical surgery • Serial patch repair • Mitral annulus • Left atrial roof

INTRODUCTION

Primary cardiac sarcomas are markedly rare (autopsy incidence of 0.001–0.03%) and the prognoses are very poor [1]. One type is intimal sarcoma, which was recently classified as undifferentiated pleomorphic sarcoma [2]. Microscopically, this tumour is composed of pleomorphic spindled or epithelioid cells with no areas of specific differentiation. Immunohistochemically, murine double minute 2 gene amplification is characteristic of cardiac intimal sarcoma [3]. Cases diagnosed by such pathological features have increased in the last decade.

CASE REPORT

A 48-year-old woman with no history of malignancy was introduced to us because of rapidly progressive heart failure. Echocardiography demonstrated a mitral valve tumour causing severe mitral stenosis (Video 1A). Chest computed tomography revealed a lobulated mass obstructing the mitral annulus. No notable metastasis or embolus was observed on systemic examinations.

Under cardiac arrest with total cardiopulmonary bypass, we obtained a good field of vision in the left atrium via a superior transseptal approach (Video 2). A tumour occupied almost the entire mitral valve and infiltrated the neighbouring left atrial intima (Fig. 1A). The anterior mitral annulus was dissected from the aortic root and the entire tumour was completely resected in one 60 × 43 × 15-mm block (Fig. 2A). There was no residual lesion macroscopically and the rapid pathological assessment strongly suggested myxoma. Therefore, we advanced the operation without additional tissue resection. Nine mattress 2-0 polyester sutures were placed along the preserved posterior annulus. The anterior annulus was reconstructed with a bovine pericardium using 6 mattress 2-0 polyester sutures placed within the...
preserved aorto-mitral curtain (Fig. 1B). The first 2 sutures were placed around the posteromedial trigone. The next 3 sutures were placed along the aortic annulus of the commissure between the left and non-coronary cusps, transmurally from the inside of the aortic root via an aortotomy. The last suture was placed in the anterolateral trigone. A 25-mm mechanical prosthesis was then implanted in the supra-annular position of the reconstructed mitral annulus. Lastly, the faulty left atrial roof was serially repaired with the bovine pericardium, which spread from the anterior mitral annulus (Fig. 1C).

The patient recovered well without complications. Postoperative echocardiography demonstrated that the replaced mitral valve worked well and that the preserved aortic valve maintained acceptable structure and function (Video 1B). However, the final pathological diagnosis was malignant intimal sarcoma. Histological examination demonstrated atypical, pleomorphic spindle cells with myxoid matrix (Fig. 2B), and fluorescence in situ hybridization study was positive for murine double minute 2 gene amplification. As the diagnosis was concluded 6 months after surgery, no adjuvant therapies were performed. The patient remains alive without recurrence for more than 1 and a half years postoperatively.

**DISCUSSION**

Reconstruction is an essential technique in radical surgery for cardiac tumours. Indeed, we reconstructed the anterior mitral annulus and left atrial roof, which were lost during tumour resection. Firm fibrous tissues of the aorto-mitral curtain, including both trigones, were preserved in addition to the aortic root and valve. By fixing the single-folded rim of the bovine pericardium to the aorto-mitral curtain, we reconstructed the anterior mitral annulus, as mentioned above. The posteromedial and

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**Figure 1:** Schema of the heart before surgery (A). Schema of the mitral annulus under reconstruction (B). Schema of the heart after surgery (C). LV: left ventricle; LA: left atrium; RV: right ventricle.
anterolateral trigones were sufficiently strong to anchor the new annulus. As the part between them had a poor appearance, the aortic annulus hiding behind in the aortic root was adopted as the underlay. At the commissure between the left and non-coronary cusps, a mattress suture was placed along the aortic annulus below the leaflets. As for the neighbouring regions, a suture was placed along the aortic annulus above each leaflet (Fig. 1B). This technique was useful for preserving the structure and function of the aortic valve, and for securing the mitral annulus. Furthermore, the left atrial roof was serially repaired with the bovine pericardium, which spread from the anterior mitral annulus. This material is sufficiently strong and flexible, although there are concerns about its durability due to degeneration. To our knowledge, this is the first report of this technique.

CONCLUSION

We performed radical surgery for a mitral valve intimal sarcoma. Serial patch repair of the anterior mitral annulus and left atrial roof with the bovine pericardium was a useful technique.

Conflict of interest: none declared.

Reviewer information

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