Clinical Features and Mid -Term and Long -Term Outcomes of Surgical Treatment of 8 Patients with Primary Ventricular Tumors

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OBJECTIVE To summarize the clinical features and surgical treatment of primary ventricular tumors.

METHODS Eight patients with primary ventricular tumor, aged 3 to 52 years, underwent surgical treatment. There were 6 males and 2 females. The pathological diagnoses were as follows: multiple left ventricular myxomas in 2 cases; left ventricular rhabdomyoma, fibroma and malignant neurolemmoma in 1 case for each; right ventricular myxoma and malignant neurolemmoma in 1 case for each; intraseptal fibroma in 1 case. The operations were performed through median sternotomy with moderate hypothermic cardiopulmonary bypass in 7 cases; via left anterolateral thoracotomy without extracorporeal circulation in 1 case. Tumors were totally removed in 7 cases and subtotally resected in 1 case.

RESULTS Cardiac arrest after anaesthetization occurred in 1 case with postoperative coma for 10 days. One case died of massive gastro-intestinal hemorrhage postoperatively. Seven cases survived, During a follow -up period of 1 to 21 years, there was no recurrence or metastasis in the 6 cases who received complete tumor resection including 2 cases with malignant tumor. One case of partial tumor removal had a mild heart murmur without tumor progression. All patients were asymptomatic with cardiac function grade I .

CONCLUSION Primary ventricular tumors showed diversity in their histological characteristics. The mid - and long -term outcomes of surgical treatment for primary ventricular tumors appear to be satisfactory.

KEY WORDS: ventricular tumor, heart tumor, surgical treatment.

Primary cardiac tumors are rare, and ventricular neoplasms are even rarer. In the present study, the clinical features and mid- and long -term outcome following surgical treatment of 8 patients with primary ventricular tumors are reported.

MATERIALS AND METHODS

Clinical Materials

From October 1981 to July 2001, 8 patients with a primary ventricular tumor underwent surgical treatment. Of the 8 cases, 6 were male, and 2 were female, aged 3 to 52 years. Six were under 30. The interval between onset of symptoms and diagnosis ranged from 3 months to 14 years. Main clinical manifestations were exertional palpitation and

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dyspnea in 7 cases, syncope in 3, a history of left atrial myxoma resection in 1, and a history of multiple pulmonary embolism in 1. The abnormalities found on physical examination were systolic murmur of grade III–IV at the 3rd and 4th left intercostal space in 3 cases, systolic murmur of grade II–III at the apex in 2, both diastolic and systolic murmurs at the apex in 1, and right-sided hemiparesis in 1. An electrocardiogram (ECG) showed left ventricular hypertrophy in 2 cases, left ventricular hypertrophic strain in 1, right ventricular hypertrophy in 2, ventricular premature beat in 2. A chest radiograph was normal in 2 cases, cardiomegaly in 2 with a cardiothoracic ratio ranging from 0.54 to 0.57 and a prominent bulge at the left cardiac border in 1. Echocardiography demonstrated ventricular tumors in 7 cases and a misdiagnosed extracavitary tumor on the posterolateral wall of the left ventricle in 1. Chest computed tomography (CT) and heart magnetic resonance imaging (MRI) were performed in 3 patients, revealing the exact size and location of the ventricular tumors.

Pathological Diagnoses

Of the 8 patients with ventricular tumors, the pathological feature was intra septal fibroma with right ventricular outflow tract stenosis in 1 case, intra-mural rhabdomyoma of the left ventricular wall with ventricular septal defect in 1, fibroma in the anterior wall of the left ventricular apex with displacement of the left anterior descending coronary artery in 1, extracavitary malignant neurolemmoma on the posterolateral wall of the left ventricle in 1, intracavitary malignant neurolemmoma of the right ventricular outflow tract in 1, right ventricular myxoma in 1, left ventricular multiple myxomas in 2, one of whom had 7 multifocal, multiple-chamber myxomas in the right atrium, left atrium and left ventricle. Tumors in the 8 cases measured from 1 cm to 6 cm in diameter.

Surgical Techniques

Operations were performed through median sternotomy with moderate hypothermic cardiopulmonary bypass in 7 patients with intracavitary and intramural ventricular tumors. Of the 7 cases, all tumors with a cuff of normal endocardium or myocardium were excised in 6 cases, partially resected in 1 case with intraseptal fibroma because the tumor was huge and adjacent to the artioventricular bundle, aortic valve, and subvalvular structures. One case received an emergency operation because of cardiac arrest after anaesthetization. All myxomas in the left ventricle were removed via a left apex incision and explorations were avoided before clamping and de-clamping the ascending aorta. Associated surgical procedures included congenital ventricular septal defect repair in 1 case, ventricular septoplasty in 1, reconstruction of the right ventricular outflow tract in 1.

Extracavitary malignant neurolemmoma on the posterolateral wall of the left ventricle was resected on the beating heart without extracorporeal circulation via left anterolateral thoracotomy in 1 case. The underlying myocardium of the tumor pedicel was electrocauterized. The left ventricular cavity was not entered.

RESULTS

There was no operative death. Postoperative coma for 10 days occurred in 1 case who had emergency operation because of cardiac arrest after anaesthetization, and who then recovered completely after comprehensive therapies. One case died of massive gastro-intestinal hemorrhage 22 days after operation. Seven cases survived. During a follow-up period of 1 to 21 years, there was no recurrence or metastasis in the 6 cases receiving complete tumor resection including 2 cases with malignant tumor. One case undergoing partial tumor removal had a mild heart murmur without tumor progression. All patients were asymptomatic with cardiac function grade I.

DISCUSSION

Primary cardiac tumors are uncommon. The incidence in autopsy has been reported between 0.0017% and 0.03% [1]. Most cardiac tumors are myxomas in the left atrium [2], and ventricular neoplasms are the least frequent, accounting for only 10% of all cardiac tumors [3,4]. In our study, the prevalence of primary ventricular tumors was 23.5% of all primary cardiac tumors in our institute (8/34), which was high compared to other reports.

The majority of primary ventricular tumors are benign, showing diversity in histological types with a wide distribution in all sites of left and right ventricles. Besides intracavitary growth, the tumors can proliferate in intramural or extracavitary forms.

To our knowledge, the multifocal, multiple-chamber myxomas, huge intraseptal fibroma, and

extracavitary malignant neurolemmoma in this series were very rare [5,6]. Clinical manifestations were relevant to the location, histological origin, and size of primary ventricular tumors. Compared with atrial tumors, ventricular outflow tract obstruction, arterial embolism, syncope and arrhythmia were more frequent in ventricular tumors because of the prominent changes in ventricular movement and pressure [6,7]. In this series of 8 cases, there was a history of syncope in 3, ventricular premature beat in 2, pulmonary embolism in 1, right-sided hemiparesis in 1 and cardiac arrest after anaesthetization caused by tumor obstruction of the right ventricular outflow tract in 1.

Clinical manifestations, chest radiograph, ECG and laboratory examinations are all non-specific for the diagnosis of ventricular tumors. Echocardiography remains the most useful modality. However, the exact location, size and number of ventricular tumors may sometimes be misdiagnosed by echocardiography because the acoustic impedance of cardiac tumors is similar to that of blood or the myocardium, and the imaging field is relatively narrow [8]. CT and MRI are highly effective in detecting tumors ,and are more and more widely applied for the diagnosis of intramural or extracavitary ventricular tumors since their imaging is wider than echocardiography, and less disturbed by lung and bone tissue [9]. In this study, malignant neurilemoma on the posterolateral wall of the left ventricle was missed with echocardiography, but found with CT in 1 case. MRI helped to develop an appropriate surgical plan in 1 case with a huge intraseptal fibroma by clearly showing the relation of the tumor to adjacent tissues.

Ventricular tumors can easily cause an embolism, ventricular outflow tract obstruction and sudden death, so an operation should be performed as soon as possible once the diagnosis is established in order to obtain satisfactory clinical results[10,11]. At present, there still are arguments regarding surgical approaches and methods. In principal, the tumors should be totally resected with care to preserve the integrity of the cardiac structure and function. Small extracavitary ventricular neoplasms can be excised on a beating heart via left anterolateral thoracotomy, while big extracavitary tumors, intramural or intracavitary ventricular tumors should be removed through median sternotomy with extracorporeal circulation. Ventricular myxomas are different from atrial myxomas. Removal of ventricular myxomas with the underlying endocardium is adequate, it is not necessary to excise the full -thickness of the myocardium. To prevent embolisms, exploration and suction cannulas are avoided before clamping the ascending aorta and the cardiac chambers are carefully rinsed after tumor resection. Soma *et al.*[5] and Schuetz *et al.* [12] excised intracavitary ventricular tumors through aortic incision or the mitral valve. In this study, 7 cases with intracavitary or intramural ventricular tumors were operated via right ventricular outflow tract or left apex incision, the ventricular tumors were completely resected in all but 1 case with a huge intraseptal fibroma. We think that ventriculotomy has the advantages of good exposure, enhancing feasibility of total resection, reducing tumor metastasis and embolisms, and preventing injury to conduction bundles and valvular tissues.

The resection and ventricular closure of large malignant ventricular tumors or benign non-myxomatous ventricular tumors may be difficult and may be followed by hemodynamic decompensation because of the small ventricular chambers. Chachques *et al.*[13] and Grinda *et al.*[14] have reported ventricular reconstruction with latissimus dorsi muscle after tumor resection. Cardiomyoplasty can preserve an adequate size and shape of the ventricular cavities, allow a complete resection without recurrence, and avoid heart transplantation. In our series, a huge intraseptal fibroma, which was adjacent to artrioventricular bundles, aortic valve, and subvalvular structures, was partially resected. There were no low cardiac output syndromes, arrhythmias, or recurrences of the tumors after the operations.

Orthotopic heart transplantation could be performed in patients with an unresectable ventricular tumor and hemodynamic decompensation. The mid- and long-term outcomes of transplantation are good [15,16]. Although chemotherapy and radiotherapy are commonly applied for malignant tumors of other organs and histological origins, their use for patients with malignant heart tumors is still controversial because most heart tumors are not sensitive to chemotherapy or radiotherapy [13,17]. In this study, underlying myocardium of the tumor pedicel was electrocauterized after the tumor was excised in 1 case with extracavitary malignant neurolemmoma to possibly kill residual tumor cells. There was no recurrence or metastasis during a follow-up period of 8 years, which implies that eletrocauterization could be a simple and feasible adjuvant therapy for some extracavitary heart tumors.

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