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Primary cardiac myxomas: report of 28 cases and review the literature

K. Durgut, R. Onoglu, N. Gormus

Department of Cardiovascular Surgery, Selcuk University, Meran Medical School, Konya

Summary

Purpose: *Although primary cardiac tumors are rare, cardiac myxoma is the most common seen intracardiac tumor. This study summarizes our surgical experience with cardiac myxomas.*

Methods: *Twenty-eight cases of cardiac myxoma that were surgically treated in our center between January 1990 and March 2010 were retrospectively reviewed. In all patients the New York Heart Association (NYHA) functional classification was used to assess the functional status, and C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) levels were measured before and after surgery.*

Results: *Nineteen patients were female and 9 male. The mean age was 53.3 ± 12.7 years (range 4-73). The most common symptom was dyspnea (86%). Two patients presented with peripheral embolism. In preoperative assessment of functional status, 24 of the patients were in NYHA Class II, 2 of them were in Class I and the other 2 patients were in Class III. All patients were in NYHA class I in the early postoperative period up to long-term follow up. The mean preoperative CRP and ESR levels were 47.3±14.6 mg/l (range 30-79) and 52.8 ± 21.4 mm/h (range 28-82), respectively. One month after surgery, CRP and ESR levels returned back to normal values. Postoperative CRP and ESR levels were 8.4±2.6 mg/l (range 1-12) and 7.9±4.8 mm/h (range 3-18), respectively.*

Conclusion: *Myxomas should be considered in all patients with embolism. After surgical resection of cardiac myxomas, the functional status of patients improve and CRP and ESR levels become normal.*

Key words: C-reactive protein, heart neoplasms, myxoma

Introduction

Primary cardiac tumors are rare and myxomas account for 45% of them. Myxomas are benign mesenchymal tumors, and 75-85% of them are located within the left atrial cavity [1,2] Symptoms are variable and nonspecific, and their occurrence depends on size, location and mobility of the tumor. However, 10% of the patients may be completely asymptomatic [3,4]. Diagnosis can be achieved by transthoracic echocardiographic (TTE) evaluation in 95.2% of the cases [5]. Surgical excision of myxoma must be performed [6,7]. After a sufficient excision, recurrence is rare [8]. In this report, we summarize our 20-year experience and review the literature about cardiac myxomas.

Methods

Patients with primary cardiac myxoma were identified by searching the cardiovascular surgery records. Between January 1990 and March 2010, 28 patients with cardiac myxoma were operated at our institute. The NYHA functional classification was used to assess the functional status of all patients before and after surgery. In all patients, laboratory tests including serum CRP and ESR were measured at the time of diagnosis and one month after surgery. TTE for diagnosis was the predominant imaging modality used in all patients. Preoperative transesophageal echocardiography (TOE) was done to confirm valvular relationship of myxoma. Intraoperative TOE was planned in need of valve repair. All patients over 40 years of age underwent a coronary angiogram to rule out coronary disease. All patients and their first degree family members were examined physically and by echocardiography for Carney complex.

In all cases, median sternotomy and cardiopulmonary bypass with moderate hypothermia was used. Myxomas were resected *en bloc* with their pedicle or with a wide cuff of normal tissue. All cardiac chambers

Correspondence to: Rasit Onoglu, MD. Selcuk University, Meram Medical School, Department of Cardiovascular Surgery, Akyokus, Konya 42080, Turkey. Tel: +90 3322236861, Fax: +90 3322236842, E-mail: drrasit@hotmail.com

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were inspected to avoid missing any tumor emboli or an occasional multicentric lesion. In right-sided myxomas, right atriotomy was performed, while in left atrial myxomas a transseptal approach after right atriotomy was used. The surgically created atrial septal defects were repaired directly or with a pericardial patch. The resected materials were examined histologically for confirmation of myxoma diagnosis.

Postoperative TTE evaluation was performed routinely to detect residual tumor, atrial septal defect and valvular functions before discharge and recurrent myxoma in the follow up period.

Results

Of our patients, 19 (67%) were female and 9 male. Their mean age was 53.3 ± 12.7 (range 24-73) years. The most common symptoms were dyspnea (86%), fever (71%), palpitation (68%), weight loss (61%), and syncope (46%). In the preoperative assessment of functional status, 24 patients were in NYHA Class II, 2 were in Class I and the other 2 were in Class III. All patients were in NYHA class I in the early postoperative period and during long-term follow up.

The mean preoperative CRP and ESR levels were 47.3 ±14.6 mg/l (range 30-79) and 52.8 ± 21.4 mm/h (range 28-82), respectively. One month after surgery, CRP and ESR levels reduced and returned back to normal values (8.4 ± 2.6 mg/l; range 2-12, and 7.9±4.8 mm/h; range 3-18, respectively).

First degree family members of all patients were 113 people. All of them were examined physically and by echocardiography for Carney complex but no one was found to have it.

Two patients presented with peripheral embolism. Embolectomy was immediately performed to shorten the duration of the limb ischemia before investigating the origin of the thrombus. Intraoperative macroscopic examination of the specimen led us to suspect the presence of tumoral embolism. After surgery, TTE was performed and confirmed heart myxoma.

In a patient with left atrial myxoma, a huge mass protruded into the left ventricle and mimicked mitral stenosis on TTE. TOE was not used routinely. Preoperative TOE was done in only 3 patients to confirm valvular relationship of myxoma. After De Vega annuloplasty of tricuspid valve, TOE was used for the same 3 patients and no postoperative regurgitation was detected.

Coronary angiogram was performed in 23 patients and 2 of them were found to have coronary artery disease. One was a 68-year-old male who was admitted to the cardiology department complaining of angina pectoris; myxoma was diagnosed incidentally on TTE. He had 95% proximal stenosis of the left anterior descending coronary artery and underwent coronary revascularization using left internal mammary artery graft after resection of the myxoma by transseptal approach. The second patient also had 85% distal stenosis of the circumflex coronary artery, which could not be revascularized because the artery diameter was less than 1 mm.

The duration of cardiopulmonary bypass was 83±22 min (range 42-125), and the cross clamp time 43±28 min (range 12-83). Transseptal approach was used in 19 patients. Right atriotomy was performed in 7 patients. Biatrial approach was needed in only 2 patients. In all patients, the location of myxomas coincided with the preoperative echocardiographic evaluation. After excision of myxoma, pericardial patch closure of the newly created atrial septal defect was needed in 2 patients. De Vega annuloplasty was performed in 3 patients, but no mitral repair was needed in our cases.

The mean mechanical ventilation time was 13.0 ± 6.8 h (range 3-24). The mean ICU stay was 2.9±1.3 days (range 1-4). The mean hospital stay was 7.5±2.2 days (range 4-11). All patients were discharged from hospital uneventfully and included in our follow up programme. The mean follow up period was 12 ± 7.4 years (range 6 months - 20 years). In our cases, no recurrence and no mortality were seen in early and late postoperative periods.

Histological examination confirmed myxoma in all patients.

Discussion

Primary cardiac tumors include benign and malignant neoplasms that arise within the cardiac chambers or myocardium. Myxomas are the most common cardiac neoplasm, accounting for 50% of all benign cardiac tumors [6]. Cardiac myxoma is a rare benign tumor with an estimated incidence of 0.5 per million population per year [8]. Cardiac myxomas usually originate from the endocardium of the atrial septum, and 95% of them are located in the atria. They are frequently seen in females between the 3rd and 6th decades of life [8].

Carney complex is an autosomal dominant disorder described in 1985 as “the complex of myxomas, spotty pigmentation, and endocrine overactivity” [9]. Some genetic defects are found associated with Carney complex [10]. No patient was found with Carney complex in our cases by physical and echocardiographical examination, but genetic analysis was not performed in our patients.

The clinical signs and symptoms produced by car-

diac myxomas are variable and nonspecific [3]. They can cause hemodynamic symptoms by obstructing blood flow or by impairing valve function. Embolization is another major hazard of cardiac myxoma. The incidence of systemic embolization is 35% in patients with left atrial myxoma [6]. The risk of systemic embolism is higher for the gelatinous, soft and polypoid or multilobular-shaped tumors than for other variants [11]. Most embolic events related to cardiac myxomas involve the cerebral arteries, followed by peripheral and mesenteric arteries [12]. In our cases, the left atrial myxomas in patients presenting with systemic embolism also had polypoid features.

Coronary embolism caused by myxoma leading to acute coronary syndrome is extremely rare. Braun et al. reported that inferior myocardial infarction related to myxoma was commonly seen, although anterior myocardial infarction was uncommon [12]. In the patient with 95% proximal stenosis of the left anterior descending coronary artery, it was difficult to distinguish whether the stenosis was related to the underlying coronary artery disease or to embolism secondary to myxoma before surgical exposure. During the operation, discovering that stenosis was related to atherosclerosis, we concluded that coexistence of coronary artery disease and myxoma in this patient was a matter of coincidence.

Myxomas are usually diagnosed incidentally while investigating unrelated symptoms and in many patients the diagnosis is delayed until the tumor has embolized or is large enough to obstruct the blood flow [8] TTE, TOE and more recently, magnetic resonance imaging are the most frequently used diagnostic tools for investigating intracardiac tumors. TOE has approximately 100% sensitivity for cardiac myxoma [5]. However, diagnosis can only be confirmed by histological examination of the excised tumor. Preoperative TTE was enough to show size, location and mobility of myxomas, so preoperative TOE was not needed in any of our patients. Intraoperative TOE was performed in 3 of our cases because of evaluation of tricuspid insufficiency after valve repair.

In myxoma patients, laboratory tests can show elevated levels of CRP, accelerated ESR, and anemia [6,11]. These are considered to be related to immune response reaction to the neoplasm and mechanical destruction of erythrocyte in blood flow. After excision of myxoma, these changes return back to normal values [13]. At the time of diagnosis our patients also showed elevated ESR and CRP levels. One month after surgery, these values were in the normal range.

Elective surgical removal is indicated whenever diagnosis of cardiac myxoma is made. Historically, it was considered an urgent procedure, particularly if the patient had a history of embolism or syncope, because it has been noted that 8-10% of the patients died of embolic complications while awaiting operation. However, recent experience suggests that elective operations (as opposed to urgent) have resulted in no greater mortality or morbidity [6]. Emergency surgery is only indicated in patients with severe dyspnoea and presenting a high risk of embolism including a history of embolism or large, mobile tumors on echocardiography [7]. Different approaches have been described for removing left atrial myxomas: right lateral, transseptal, transseptal biatrial and superior transseptal. In our cases of left atrial myxoma, the transseptal approach was preferred. The superior transseptal approach has been reported as an excellent method in comparison with others since it provides a clear surgical view for resecting left atrial myxomas [14].

Surgical resection is an accurate approach for the treatment of myxoma in any cavity of the heart. The important point of the operation is to achieve complete excision of myxoma and the affected area, and maximum effort should be spent to preserve the original valve structure. After resection of tumoral mass, if necessary, the atrial septal defect must be closed primarily or with pericardial patch and valve reconstruction or replacement must be performed [15]. De Vega annuloplasty was performed in 3 patients in our cases because of tricuspid valve insufficiency after excision of myxoma. In 2 patients, the atrial septal defect created after excision of myxoma was repaired using pericardial patch.

Right-sided cardiac myxomas are uncommon, and they can cause pulmonary embolism before or during surgery. In these cases, a surgical strategy must be planned to prevent pulmonary embolism. To achieve this, the pulmonary artery can be cross clamped as soon as extracorporeal circulation is initiated and a single cannula in the superior vena cava can be used until fibrillation before another is inserted into the inferior vena cava [16]. In 7 patients with right-sided myxoma these strategies were performed and no pulmonary embolism occurred.

Velicki et al. reported that, because of relatively low risk, surgical treatment was recommended for most patients with excellent postoperative prognosis [17].

Removal of atrial myxomas carries an operative mortality rate of 5% or less [3]. Kajihara et al. reported that the hospital mortality after an excision of myxomas was 2.2% [13]. In our cases, no mortality was seen in early and late postoperative period.

Long-term results after resection of myxoma are usually satisfactory. After a sufficient excision, recurrence is rare [8]. In the follow-up period, transthoracic echocardiographic evaluation is recommended for the detection of recurrent myxoma and diagnosis can be achieved in 95.2% [5]. Recurrence usually occurs with-

in 48 months after the initial resection [8]. Incomplete resection, intracardiac implantation, embolization, and a reserve of tumor precursor cells in the subendocardium are the risk factors for recurrence [3]. No recurrent myxoma was diagnosed in our cases. However, all patients are in our follow-up programme and echocardiographic evaluation is performed every year.

Conclusion

Myxomas should be considered in all patients with embolism. TTE is beneficial, easy to use and highly sensitive tool for diagnosis. We suggest the transseptal approach for removing left atrial myxomas. After surgical resection of cardiac myxomas, functional capacity of patients improve, CRP and ESR levels become normal. Long-term survival appears excellent after surgery.

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