Case report - Cardiac general

Successful surgical treatment of a right atrial myxoma complicated by pulmonary embolism

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Abstract

We report on a rare case of a 65-year-old woman who was admitted with orthopnea (NYHA class IV) and a single syncopal episode. A transthoracic echocardiography examination showed a $7 \times 6 \times 6$-cm mass located in the right atrium and perfusion lung scan showed embolization. Complete surgical removal of the right atrial myxoma and tumor embolectomy was successfully performed with deep hypothermia and circulatory arrest. Histologic investigations of the primary tumor as well as the emboly revealed benign myxoma. Six months clinical and echocardiographical follow-up showed a satisfactory exercise tolerance, a normal right atrium and a good ventricular function. Thirteen years later, the patient remains well and no recurrence could be observed.

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1. Introduction

In 1908, a right atrial myxoma was first described [1]. In the next 50 years, pulmonary embolism due to right heart myxomas were reported by the pathologists, to our knowledge the first case was described by Hermann Chiari in Vienna [2]. In 1954, Clarence Crafoord performed the first successful resection of a cardiac myxoma [3]. Primary cardiac tumors are rarely found and have an incidence of 0.3% of all open-heart operations. Among those, about 70% are myxomas, most of them in the left atrium [4]. Less than 20 similar cases with pulmonary embolism due to myxoma have been reported [5,6], some of them postmortem [7] or with postoperative mortality [8].

2. Case report

We report on a previous healthy 65-year-old woman who was admitted with orthopnea (NYHA class IV) and a single syncopal episode. Physical examination showed swollen ankles, but no signs of peripheral deep vein thrombosis. Chest X-ray revealed moderate enlargement of the cardiac silhouette with normal pulmonary fields. Standard ECG showed sinus tachycardia and an incomplete right bundle branch block. A transthoracic echocardiography (TTE) examination showed enlargement of the right chambers and a $7 \times 6$-cm mass located in the right atrium (Fig. 1) and prolapsing through the leaflets of the tricuspid valve during diastole. The mass showed a small degree of motion, but no free-floating movement suggestive of a mobile thrombus. TTE showed good left ventricular function, right ventricular function slightly reduced. A lung ventilation–perfusion scan showed absence of flow to the entire right lung. Computed tomography revealed no further information, only the tumor was seen. Left coronary angiogram revealed a good ventricular function (ejection fraction 0.70) without signs of coronary artery disease.

Urgent open-heart surgery was arranged. Cardiopulmonary bypass was established by ascending aortic and bicaval cannulation (to avoid tumor embolization) using two standard 28-Fr venous return cannulas. We performed en bloc resection of the right atrial tumor with a cuff of normal septum tissue with direct closure of the interatrial septum. Under deep hypothermia (20 °C) and circulatory arrest...
After arteriotomy of the right pulmonary artery, a 3-cm-diameter tumor fragmentation was removed (9 min). The postoperative course was uncomplicated. The patient was discharged from our hospital on the 12th day. Histologic investigations of the primary tumor, a 6 × 6 × 7-cm large, friable and gelatinous mass (Fig. 2) as well as the 2 × 3 × 2-cm large embol revealed benign myxoma with large areas of myxoid matrix and areas with hemorrhage within the tumor mass. Six months later the patient was asymptomatic. Follow-up echocardiography showed a normal right atrium and a good ventricular function (ejection fraction 0.60). Thirteen years later, the patient remains well and no recurrence could be observed.

3. Discussion

Pulmonary embolism in the majority of cases implicates embolization from thrombotic material from a vein, from the right heart chambers or from catheter tips. In the absence of risk factors or clinical evidence for thrombosis, suspicion of non-thrombotic pulmonary embolism may arise. Symptoms of cardiac tumors are often misleading although severe or catastrophic complications such as embolization, coronary occlusion, syncope as in our case, loss of consciousness of unknown etiology, infection and sudden death may occur. The signs are highly dependent on the position of the myxoma. Often, these patients demonstrate the potential for diagnostic confusion and significant delay in diagnosis, especially in the presence of extracardiac symptoms. We concluded that tumor fragmentation and embolization of a right atrial myxoma into the pulmonary arteries is a rare source of pulmonary embolism. Infrequently, intracardiac thrombus mimicking myxoma and the exact diagnosis is only established by histologic investigation of the operative specimens.

All patients with idiopathic (recurrent) pulmonary embolism should undergo early echocardiography. In the presented case, the enlargement of the right chambers in echocardiography led to diagnosis. It is well known and also from our experience that transesophageal (TEE) is superior to transthoracic echocardiography in the evaluation of right atrial tumors and TEE should be considered in patients with right atrial masses even when these tumors have been demonstrated with TTE [9]. Sensitivity is up to 100% and better than computed tomography or magnetic resonance tomography, but echocardiography is not specific for cardiac tumors. Nevertheless, one report describes fatal pulmonary embolization of the right atrial mass during TEE investigation [10]. During surgery, TEE provides useful information to secondary valve damage caused by the tumor or to detect rare biatrial or multilocular myxomas and we recommend it as a intraoperative routine during resection of cardiac masses.

We recommend bicaval cannulation to avoid tumor fragmentation. Hypothermic circulatory arrest is the method of choice and provides good view for embolectomy, sucker devices or Fogarty catheter maneuvers are not recommended.
References


