Case Report

Bilateral Atrial Myxoma: A Case Report

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Among the rare cardiac tumors, myxoma, which is mostly located in the left atrium, is the most common type. Bilateral atrial myxoma is extremely rare, and requires urgent surgery. The authors report the case of a 34-year-old male, who presented with one month of right hemiparesis and aphasia and subsequently diagnosed with bilateral atrial myxoma based on transthoracic echocardiography. An urgent operation for intra-cardiac tumor removal was performed with the biatrial approach. Once a diagnosis of myxoma has been made, an urgent operation for tumor removal is necessary due to the risk of serious complications, including sudden death from normal blood flow obstruction.

Keywords: *Cardiac tumor, Myxoma*

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Myxoma is the commonest cardiac tumor. It is two to three times more prevalent in women than in men, and it is more common in older adults than in children. Eighty percent of myxomas are in the left atrium, and they usually arise in the atrial septum[1]. Bilateral atrial myxoma is extremely rare. A classical presenting triad is embolization, heart failure due to blood flow obstruction, and other constitutional symptoms.

Case Report

A 34-year-old gentleman was hospitalized with right hemiparesis and aphasia while resting at home. Computed tomography of the brain revealed a large infarction at the left cerebral hemisphere. His cardiovascular physical examination was normal. Further investigation into the source of the emboli was performed, including transthoracic echocardiography, which revealed a large, inhomogeneous, mobile mass arising from the interatrial septum to both the left and right atria that somersaulted through the right ventricle, and there was mild tricuspid valve regurgitation with no visible intra-cardiac thrombus (Fig. 1). Bilateral atrial myxoma was diagnosed, and the decision was made to perform urgent intra-cardiac tumor removal.

Full median sternotomy and opening of the pericardium were performed, and the heart was cannulated for cardiopulmonary bypass. Double direct venous cannulation of the superior and inferior vena cava was generally used. The core body temperature was reduced to 30°C, and snuggling of both vena cavae was performed for right atrium isolation. The ascending aorta was cross-clamped, cardioplegic solution was delivered through the aortic root, and right and left atrial incisions were made to explore both atriums.

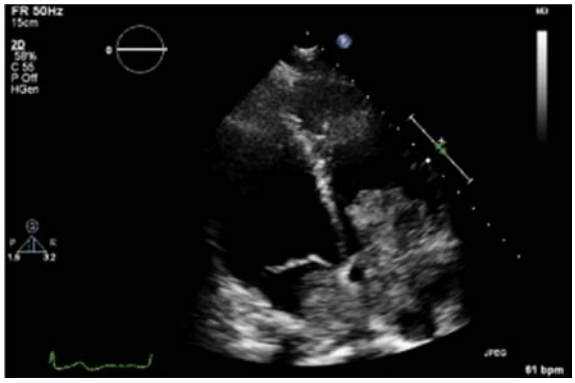

Fig. 1 Inhomogeneous mobile mass attached to the interatrial septum in both the right and left atria with somersaulting of the mass through the right ventricle.

A large cerebriform pedunculated tumor almost entirely filled the right atrium. The tumor was arising from the same area of the interatrial septum to a broad base that was attached to the dorsal free wall of the right atrium on the inferolateral aspect, measuring 6.5x6x2 cm; on the left side, it was attached to the

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anterior wall of the left atrium, measuring 1.5x1x0.7 cm. The stalk of the septum was 1 cm in length and 0.5 cm in diameter (Fig. 2).

A circular septal incision was made around the tumor origin after which en bloc resection of the tumor was performed with an atrial septum (Fig. 3). The defect in the atrial septum was reconstructed with a Dacron patch. The atriotomy was closed, the aortic cross-clamp was removed, and expulsion of air was performed via the aortic root vent. When cardiac contraction resumed, the heart was allowed to fill while suction was maintained on the aortic root. When the patient was fully warm and cardiac function was restored, cardiopulmonary bypass was discontinued. A pacing wire was placed on the right ventricle wall after which the pericardium, sternum and chest wall were closed. Immediate post-operative transesophageal echocardiography showed no residual mass, but mild tricuspid valve regurgitation remained.

On the second day after surgery, the patient was transferred from the intensive care unit to the ward. He spent some time in the postoperative rehabilitation program, then discharged seven days after surgery. Later, the pathological report confirmed the diagnosis of bilateral atrial myxoma (Fig. 4).

Discussion

Primary cardiac tumors are rarely found in the general population. Lam et al found that the incidence of primary cardiac tumor was 0.056% in 12,485 autopsies from 1972-1991[2]. Among those tumors, myxoma accounted for more than 50% of benign cardiac tumors, and it mainly affects women between 30 and 60 years of age[3]. The location of cardiac myxoma was mostly in the left atrium (82.5%), followed by the right atrium (15.5%)[4]. The most common presentation involves symptoms of mitral valve obstruction with cardiac failure or malaise (67%), and constitutional symptoms and embolism are found in 34% and 29%, respectively[5]. Bilateral atrial myxoma is extremely rare, and its first successful removal was reported in 1967 by Yipintsoi et al[6]; since then, few cases have been reported. The mortality after removal of atrial myxomas is less than 5%[5] and recurrence is rare, occurring in approximately 2% of cases[7]. Atrial myxoma may be

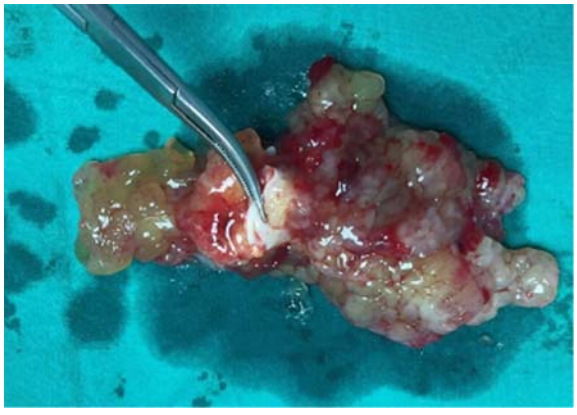

Fig. 2 Specimen of the bilateral atrial myxoma; part of the interatrial septum is at the tip of the clamp.

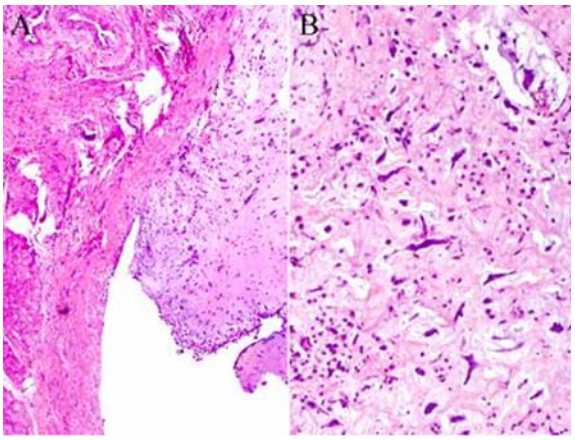

Fig. 3 Microscopic findings revealed the following: (A) Interface between the atrial septum and the stalk of the myxoma (H&E X40) and (B) Stellate and globular myxoma cells in abundant myxoid matrix (H&E 600X).

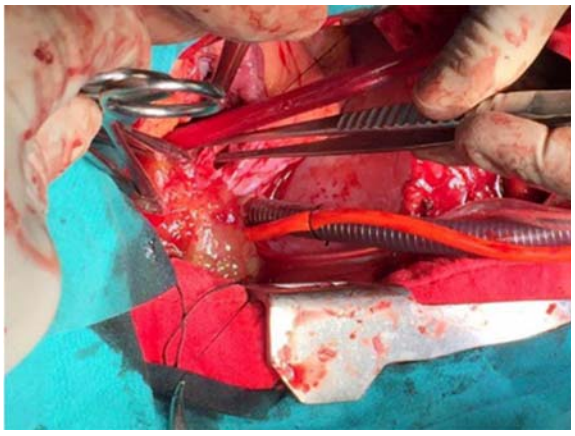

Fig. 4 En bloc resection of the bilateral atrial myxoma; the atrial septum that was resected before it was reconstructed with a Dacron patch is located at the tip of the forcep.

multicentric (within a single chamber) or biatrial. Most biatrial myxoma has the same attachment area for two stalks to the opposite site of the same interatrial septum[8].

Most cases of atrial myxoma are sporadic with unknown etiology[9], but familial cardiac myxomas are found in nearly 7%[10] of patients with autosomal dominant transmission. It may present as part of an associated syndrome, including Carney complex (a frame-shift mutation in exon 2 in a heterozygous fashion in the causative gene of the Carney complex, protein kinase A regulatory subunit 1 alpha)[11], LAMB syndrome (Lentigines, Atrial myxomas, Mucocutaneous myxomas, and Blue nevi), NAME syndrome (Nevi, Atrial myxoma, Myxoid neurofibroma, and Ephelides) and nonsyndromic familial cardiac myxoma[12]. It is important to be aware of the familial type of cardiac myxoma in cases of bilateral atrial, multifocal or recurrent myxoma. Myxomas are vascular tumors, and immunohistochemistry analyses of the cytoplasm of tumor cells have revealed the presence of vascular endothelial growth factor (VEGF) and its receptors[13]. Myxoma cell proliferation is enhanced by the addition of VEGF in a dose-dependent manner and is inhibited by the addition of a neutralizing VEGF antibody[14]. Furthermore, the levels of both VEGF and interleukin-6, an inflammatory cytokine, are raised[15], and these molecules may potentially play a role in constitutional symptoms, such as fever, weight loss, arthralgias and Raynard phenomenon. Cardiac myxoma may be neovascularized by a branch of the coronary artery[16], but pre-operative coronary angiogram currently has no role in its further management.

Although atrial myxoma is rare, it should be considered as one of the differential diagnoses because it is a life-threatening condition. Patients who present with unexplained symptoms of low cardiac output or ischemic stroke should undergo a careful physical examination, including echocardiography. Once atrial myxoma is diagnosed, urgent surgery is indicated, as this condition may cause an obstruction of the normal cardiac blood flow. Patients should be checked for recurrence even though the recurrence rate is low. The protocol for long-term follow-up with consistent echocardiogram should be scheduled after surgery.

What this study adds ?

Bilateral atrial myxoma is extremely rare, and urgent surgery is indicated because this tumor may obstruct the blood flow. Therefore, echocardiography follow-up is recommended to check for recurrence.

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Potential conflicts of interest

None.

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เนื้องอกของหัวใจชนิด myxoma ทั้งสองฝั่งของหัวใจห้องบน: รายงานผู้ป่วย

อรรถภูมิ สู่สุภอรรถ, กำพู ฟูเฟื่องมงคลกิจ

ในกลุ่มเนื้องอกของหัวใจที่พบได้น้อย เนื้องอกของหัวใจชนิด myxoma สามารถพบได้มากที่สุด และมักพบอยู่ภายในหัวใจห้องซ้ายบน การพบเนื้องอกชนิดนี้อยู่ทั้งสองฝั่งของหัวใจห้องบนพบได้ยากมาก และเป็นข้อบ่งชี้ในการผ่าตัดเร่งด่วน ผู้นิพนธ์รายงานผู้ป่วยชาย อายุ 34 ปี ซึ่งมาด้วยอาการแขนขาซีกขวาอ่อนแรงและไม่สามารถพูดโต้ตอบได้ 1 เดือนก่อนมาโรงพยาบาล ซึ่งภายหลังตรวจพบเนื้องอกภายในสองฝั่งของหัวใจห้องบนจากภาพคลื่นเสียงสะท้อนของหัวใจ ก่อนจะได้รับการผ่าตัดเพื่อนำก้อนเนื้องอกออกโดยการผ่าตัดแบบเปิดหัวใจห้องบนทั้งสองฝั่ง ผู้ป่วยหากได้รับการวินิจฉัยภาวะนี้แล้วควรได้รับผ่าตัดเร่งด่วน เนื่องจากอาจเกิดภาวะแทรกซ้อนที่ร้ายแรงถึงแก่ชีวิต เนื่องจากการอุดกลั้นของทางเดินเลือดปกติในหัวใจ