An Exceptional Cause of Progressive Dyspnoea in a Renal Transplant Recipient: Hemangioma of the Mitral Valve

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Abstract
Primary cardiac hemangioma is a very rare benign vascular tumor, with valvular hemangiomas being even less frequent as valves are generally avascular structures. We present the first case of mitral valve hemangioma in a renal transplant recipient. Patient presented with progressive dyspnea. Transesophageal echocardiogram (TEE) demonstrated a 0.8x0.9-cm pedunculated tumor mass on the posterior leaflet of the mitral valve. Coronary angiography identified a small artery which filled from the circumflex artery and fed the tumor. The tumor was surgically removed. Histopathological examination revealed a hemangioma. The postoperative course was uneventful with stable graft function.

Introduction
Primary cardiac hemangioma is a very rare benign vascular tumor. Valvular hemangiomas are even less frequent as the valves are generally avascular structures [1, 2]. They can be found in any age group, but occur most commonly in childhood. To our knowledge, less than 10 cases of mitral valve hemangiomas have been documented in the literature [3-11].

We present the first case of mitral valve hemangioma in a renal transplant recipient.
Case report

A 49-year-old woman with progressive dyspnea was admitted to the cardiology ward. Her medical history included chronic pyelonephritis with development of the end-stage renal disease. She had been treated with hemodialysis for 9 years, and in 2006 received a renal transplant from a deceased donor. Graft function was excellent with serum creatinine 89 µmol/L. Five years after transplantation she presented with symptoms of progressive dyspnea and chest tightness that worsened especially during intensive physical activity. Physical examination revealed a normal heart rhythm with clear sounds and a systolic murmur over the mitral valve. Laboratory tests including routine hematology, biochemistry and coagulation were all within the normal range. She was afebrile. A transesophageal echocardiogram (TEE) demonstrated a 0.8x0.9-cm pedunculated tumor mass on the posterior leaflet of the mitral valve (Fig. 1) which was not present on the heart ultrasound two years before. Coronary angiography revealed a small artery filling from the circumflex artery and feeding the tumor (Fig. 2). Cardiac surgery was indicated.

A median sternotomy was performed. The left atrium was opened and a 0.8x0.6-cm solid red mass was excised from the posterior leaflet of the mitral valve. The operation lasted 44 minutes, with 18 minutes of cardiac ischaemia. Histopathological examination revealed hemangioma (Fig. 3).

The postoperative course was uneventful. Graft function remained stable and the patient was discharged from hospital 7 days after the surgery in good condition, with complete regression of cardiac symptoms. The control heart ultrasound revealed a functionally competent mitral valve with no residual tumor mass. After 8 months she is still doing well, with no signs of hemangioma recurrence, with normal cardiac and renal function.

Discussion

Solid organ transplantation with chronic immunosuppression is associated with an increased risk of malignacies [12, 13]. There are national registries reporting increased incidence of malignancies in renal transplant recipients [14-19]. However, until now there is no evidence suggesting an increased risk of heart hemangioma in general or heart valve
hemangioma in patients with immunosuppressive therapy due to solid organ transplantation. There is only one isolated report of an accidental finding of aortic valve hemangioma found in a renal transplant recipient [20]. Our case would be the second case of valve hemangioma following renal transplantation and solid organ transplantation in general and the first case of mitral valve hemangioma in a renal transplant recipient described in the literature.

Primary cardiac tumors are rare findings [1]. Their incidence found on autopsy was 1.7 per 100,000 cases, with about 75% being benign [21]. Of these, cardiac hemangiomas are extremely rare tumors which may involve endocardium, myocardium or epicardium [2], and account for less than 3% of all primary cardiac tumors [22] and 5-10% of all benign cardiac tumors [21]. Valvular hemangiomas are even less frequent as cardiac valves are predominantly avascular structures.

Based on our review of the literature there is no data on the growth dynamic of mitral valve hemangioma. All reported cases were diagnosed after symptoms occurred with no prior evidence of any echocardiography finding or/and no data regarding the time between the last echocardiography finding and diagnosis of mitral valve hemangioma. Our case shows relatively quick growth of the hemangioma, hence it was absent in an echocardiogram performed two years before the presentation. The growth hemodynamics of heart hemangiomas in general are unpredictable ranging from dormancy, to accelerated growth, to spontaneous regression/involution [23, 24]. Lesions are typically discovered incidentally during routine diagnostic procedures [22] as the majority of patients remain asymptomatic. Their clinical presentation varies according to the size, location and mobility of the tumor. However, the majority of lesions are incidental finding. The presenting symptoms of heart valve hemangiomas may include palpitations and syncope, exertional dyspnea, heart failure due to hemodynamic disturbances, and atypical chest pain. [3, 6, 9, 10, 25-27]. In addition to these symptoms, the presenting symptoms of a cardiac hemangioma localized elsewhere than on the valves may be arrhythmias [28, 29], embolic incidents [30-32], sudden cardiac death [33], pericardial effusion [34, 35] and heart tamponade [36]. Echocardiography may raise suspicion of this rare tumor; however, it may be very challenging as less than one third of reported cases of hemangiomas were diagnosed preoperatively [37]. Coronary angiography may delineate feeding vessels to the tumor with a characteristic „tumor blush” [28, 38]. In our patient the tumor was supplied by a branch of the circumflex artery and this finding was suggestive of a vascular etiology of the tumorous formation with definitive confirmation of the benign nature of the lesion on the pathohistological finding.
Only one case of incidentally found aortal valve hemangioma in a renal transplant recipient has been reported in the literature. This was 62-year-old man who presented with syncope and dyspnea. He had chronic, degenerative aortic valve stenosis, and hemangioma was discovered during aortic valve replacement [20].

Potential life-threatening complications of valvular hemangiomas require prompt surgical intervention. Long-term prognosis is excellent after complete recovery. There is no evidence of cardiac hemangioma recurrence in the longer time follow-up period [4, 39]. Other reported cases reveal data from a shorter follow-up period such as one to two years [9, 10], or no data at all [26, 40]. No recurrences of cardiac hemangioma have been reported so far, but given the rareness of the condition and hence the uncertain prognosis, regular follow-up is probably warranted [3].

Conclusion

In conclusion, our case demonstrates that renal transplant recipients may suffer from different cardiovascular problems including very rare cardiac tumors. The diagnosis of a cardiac valve tumor should be taken into consideration in renal transplant recipients presenting with typical symptoms. Echocardiography is the most appropriate initial screening and diagnostic imaging modality. Surgical excision is recommended with regular ultrasound follow-up to discover possible tumor recurrences.

Conflict of Interests

The authors of this manuscript state that they have no conflicts of interest.

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