

Myxoma of Donor Origin in a Transplanted Heart

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Myxomas are the most common primary cardiac tumors but their presence in the transplanted heart is extremely rare. We report a case of left atrial myxoma in a patient after heart transplantation. DNA analysis confirmed a donor origin. To our knowledge, this is the first report of myxoma of donor origin in a transplanted heart. *J Heart Lung Transplant* 2007;26:865-7. Copyright © 2007 by the International Society for Heart and Lung Transplantation.

Cardiac myxomas, particularly left atrial myxomas, are the most common primary tumors of the heart. Myxoma in a transplanted heart is exceptionally rare. To our knowledge, this is the first reported case of a myxoma of donor origin in a transplanted heart.

CASE REPORT

A 51-year-old man with a history of advanced heart failure due to ischemic cardiomyopathy underwent orthotopic heart transplantation at our institution in August 1998. The heart donor was a 26-year-old man with isolated cranial trauma. The early post-transplantation period was uneventful. The immunosuppressive regimen consisted of cyclosporine, azathioprine and prednisone. Only a single episode of acute rejection, which resolved after corticosteroid pulse therapy, was observed within the first year after the procedure. In February 2002, prednisone administration was discontinued. There was an unsuccessful attempt to replace azathioprine with mycophenolate mofetil due to severe gastrointestinal discomfort. In March 2006, cyclosporine was switched to tacrolimus for better control of lipid metabolism. During the follow-up period, systolic function of the graft remained normal.

In August 2006, 8 years after the transplant, routine transthoracic echocardiography revealed an oval-shaped, pedunculated mass in the left atrium. Subsequent transesophageal echocardiography demonstrated the mass in

detail and showed an attachment to the roof of the left atrium (Figure 1a). The appearance of a tumor suggested a myxoma, with a size that reached 4.8 cm × 2.5 cm × 2.0 cm. Multi-detector computerized tomographic angiography supported this diagnosis (Figure 1b). Coronary angiography showed non-significant stenoses of the coronary arteries.

The patient was clinically entirely asymptomatic and was scheduled for surgical extirpation of the tumor. The procedure was performed in September 2006 using extracorporeal circulation. The left atrium was incised anteriorly to expose the mass. The pedicle, measuring 2 cm in length, originated from the area of the previous atrial suture line. The whole tumor, measuring 6.0 cm × 4.0 cm, was then excised. The diagnosis of myxoma was confirmed (Figure 2a and b). The post-operative course was complicated by the development of Type A aortic dissection. The dissection was discovered post-operatively and spread to both iliac arteries, resulting in impaired perfusion of visceral organs. Despite early re-operation with revision and suture of the intimal flap in the aortic arch, the patient subsequently died of hepatorenal failure.

To study the origin of the myxoma, DNA was isolated from whole venous blood and from tissue specimens obtained from the tumor according to the Miller et al.¹ Two independent polymerase chain reactions (PCRs) were performed to distinguish the genetic background of the tumor tissue^{2,3} (Figure 3). The patient was heterozygotic for the insertion/deletion variant within the angiotensin-converting enzyme gene (Figure 3, see 2 at top; bands: A, insertion; B, deletion). PCR product from the tumor tissue lacked the insertion band A (Figure 3, see 3). A second PCR with the primers specific for the insertion confirmed the presence of an insertion allele in the patient blood (Figure 3, see 5, band C), whereas the allele was missing in the tumor tissue (Figure 3, see 6). This result confirmed a donor origin of the myxoma.

DISCUSSION

In a previous study, the autopsy incidence of primary cardiac tumors was found to range from 0.001% to

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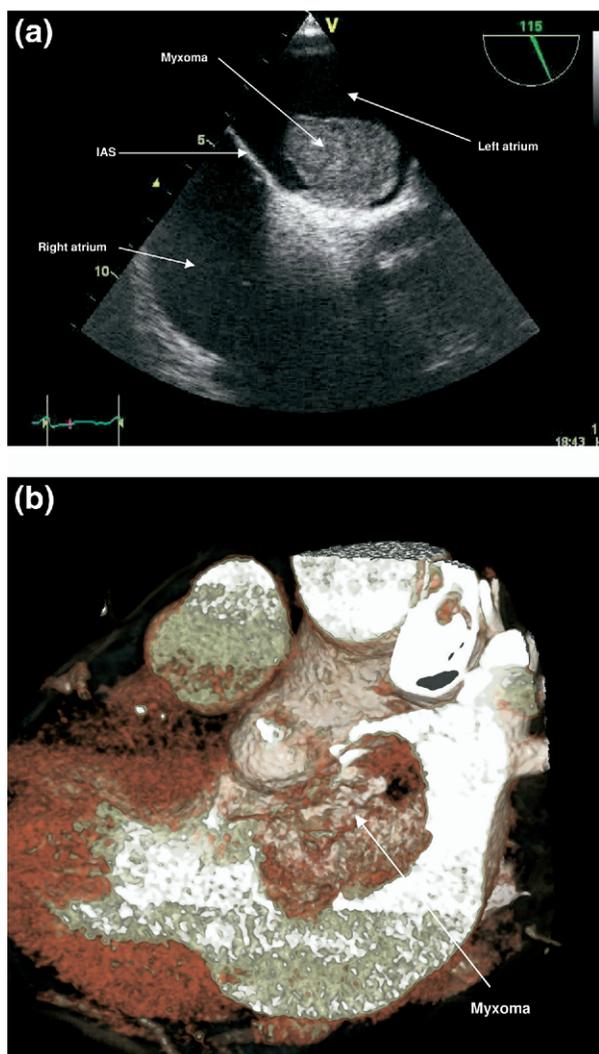


Figure 1. (a) Myxoma in the left atrium of a transplanted heart (transoesophageal echocardiogram). (b) CT scan of the myxoma.

0.030%.⁴ Seventy-five percent of these tumors are benign and nearly 50% of benign tumors are myxomas. About 75% are located in the left atrium, usually arising in the atrial fossa ovalis region. Myxomas can cause many serious complications, such as recurrent strokes, peripheral or pulmonary embolization, valve obstruction, etc. Myxomas could be familial and, in rare cases, multiple as a part of a Carney complex, which also involves lentiginos, blue naevi and fibromyxoid tumors of the skin.^{4,5} The treatment of choice for myxomas is complete surgical excision. Prognosis after surgical therapy is quite good, although there is a 5% recurrence rate.⁶

Occasionally, myxoma could be an indication for autotransplantation or orthotopic heart transplantation. The literature has reported a few cases of giant myxomas, which could not be resected completely. Therefore, the technique of cardiac autotransplantation was used.^{7,8} Goldstein et al⁹ described the case report of a

patient with an extensive, recurrent atrial myxoma and progressive mitral stenosis, which subsequently was the reason for orthotopic heart transplantation.

Myxomas are typically characterized by an ovoid, spherical shape and have a mottled appearance. Although echocardiographic and macroscopic assessment of the mass in the left atrium would seem to indicate myxoma, it may prove to be something different on pathologic assessment. For example, left atrial thrombus¹⁰ and leiomyosarcoma¹¹ have been mistakenly diagnosed as myxomas. In both cases only histologic examination could exclude the incorrect diagnosis.

The association between myxoma and immunosuppressive therapy remains a matter of discussion. On the one hand, life-long immunosuppressive therapy is necessary after organ transplantation. On the other hand, it is associated with various side effects, including an increased incidence of malignancies, with tumors of the skin, digestive tract and lungs and lymphoproliferative disease being the most frequent.¹²

There is another important issue with regard to heart transplantation—whether to accept or reject donor hearts with pre-operatively diagnosed myxoma. There are only two reports in the literature on rejection of a heart due to myxoma. In the first case, a left atrial

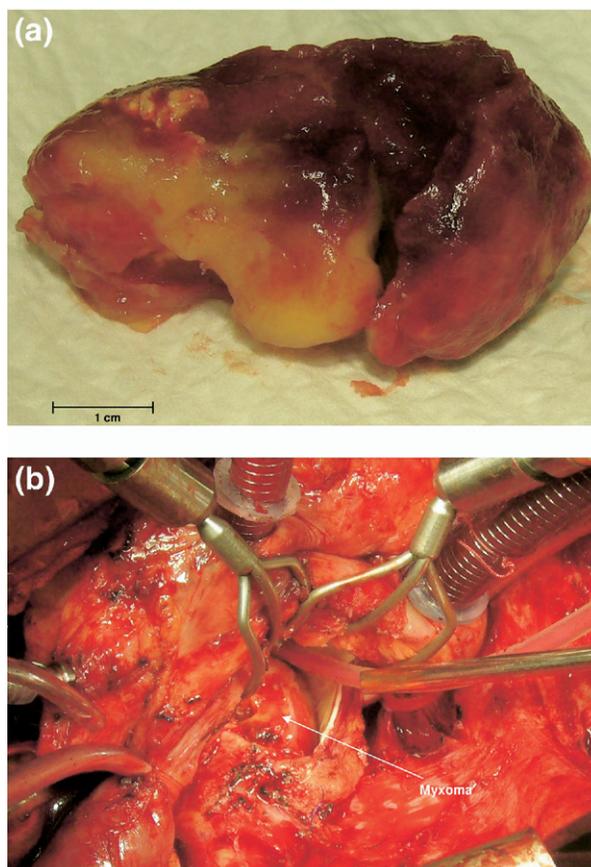


Figure 2. (a) Resected myxoma. (b) Myxoma in the left atrium.

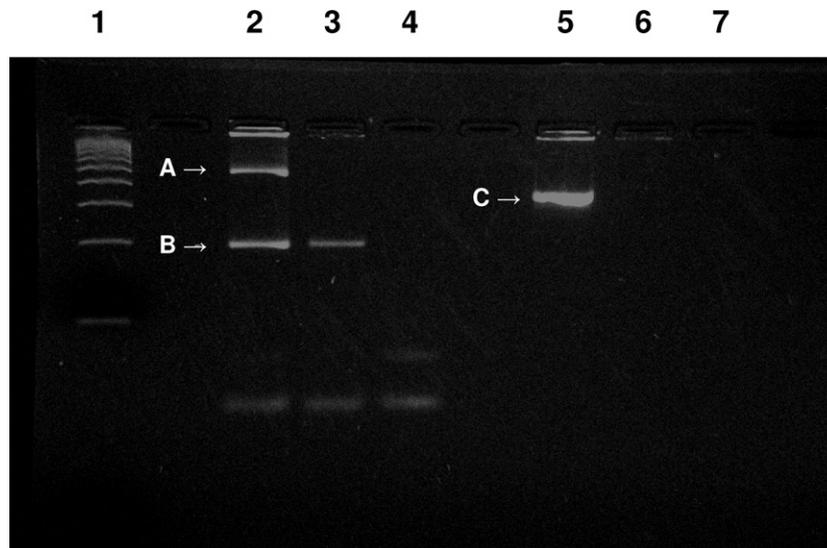


Figure 3. DNA analysis of the recipient DNA and DNA from the myxoma: 1—DNA standard; 2—genotyping reveals that the patient is heterozygotic [A + B] for the ACE insertion/deletion polymorphism; 3—in DNA from the tumor, insertion allele B is missing. Insertion-specific PCR further confirmed the presence of the insertion allele in the patient (5, C) but not in the tumor (6); 4 and 7—negative controls.

myxoma in a donor heart was revealed by echocardiography and, due to suspicion of a Carney complex, the heart was not harvested.¹³ In the second case, a pathologic lesion on the anterior leaflet of the mitral valve was found during inspection of the heart by the thoracic surgeon. Histologic examination of the lesion showed typical myxoid tissue, confirming the diagnosis of a cardiac myxoma.¹⁴ In our opinion, a donor heart with myxoma may be acceptable for urgent recipients in special cases when the myxoma appears to be extirpated easily, without connection to valves and with no signs of a Carney complex.

Finding a myxoma in a transplanted heart is exceptionally rare. To our knowledge, this is the first report of a left atrial myxoma of donor origin occurring in transplanted heart. We found another report in the literature of an atrial myxoma in a transplanted heart that originated from recipient's heart, as confirmed by DNA analysis.¹⁵ Another case of atrial myxoma was reported after bone marrow transplantation.¹⁶

Surgical extirpation is the treatment of choice for this condition. The risk associated with this procedure is relatively low, but some complications can occur. Neither tumor nor its extirpation caused the death in this case, but we cannot exclude injury of the aortic wall during cannulation as a cause of aortic dissection.

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