Case Reports

Recurrent Biatrial Myxoma in a 41-Year-Old Woman after Left Atrial Myxoma Resection

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Bilateral recurrence of atrial myxoma has been reported only a few times. We describe the case of a 41-year-old woman who had undergone left atrial myxoma resection and presented 9 years later with myxomas in both atria. The patient underwent successful resection of both masses with resolution of symptoms. We discuss the phenomenon of recurrent biatrial myxoma. ***(Tex Heart Inst J 2017;44(6):402-4)***

Key words: *Cardiac surgical procedures; heart atria/pathology; heart neoplasms/pathology/surgery; myxoma/diagnosis/genetics/surgery; neoplasm recurrence, local; neoplasms, multiple, primary/surgery; reoperation; treatment outcome*

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Cardiac myxomas are primary cardiac neoplasms composed of primitive connective tissue cells and stroma. Although benign, these intracardiac masses are often removed because they can lead to severe complications. Recurrence of myxomas is very uncommon in patients without familial predispositions. We describe a sporadic, biatrial recurrence of myxoma in a patient who had undergone resection of a left atrial myxoma.

Case Report

A 41-year-old woman with a history of left atrial myxoma presented with palpitations and dyspnea on exertion. The palpitations had begun the night before and had prompted her visit to the hospital. At age 32 years, she had had multifocal cerebral infarcts, and an echocardiogram had revealed a 5 × 4-cm myxoma in her left atrium. The mass was resected with clear margins, and the patient remained asymptomatic until one year before the current presentation, when she began experiencing intermittent palpitations, dyspnea on exertion, orthopnea, and lower-extremity swelling.

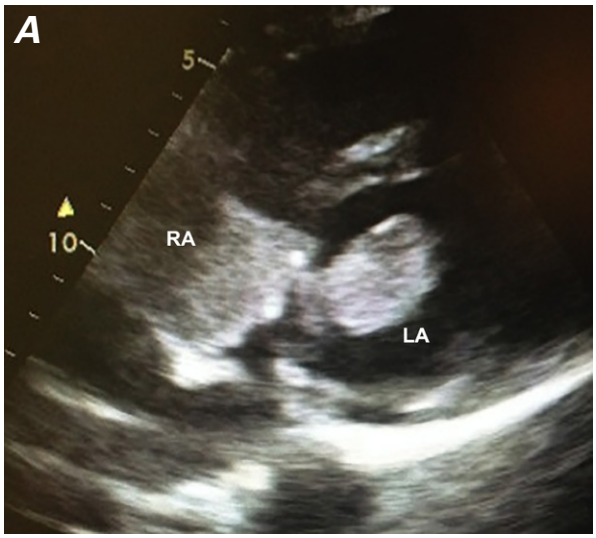
Upon presentation, the patient had atrial flutter but was otherwise hemodynamically stable. Physical examination revealed a holosystolic murmur and tumor plop, but no signs of congestive heart failure. Transthoracic echocardiograms and cardiac magnetic resonance images showed a large mass occupying almost the entire right atrium and a hypermobile left atrial lesion that prolapsed through the mitral valve during diastole (Fig. 1).

The patient was taken to the operating room for repeat median sternotomy. A right atriotomy exposed a 5 × 4 × 3-cm mass adherent to the anterolateral free wall. This mass was resected. Then, the 3 × 2.5 × 1.5-cm left atrial mass, along with part of the atrial septum to which the tumor was attached, was removed through a transseptal approach. A closed foramen ovale was confirmed, the septum was repaired, and the right atrium was closed. Both surgical specimens were sent for pathologic evaluation, and the diagnosis of cardiac myxoma was verified (Fig. 2).

The patient tolerated the procedure well and had no noteworthy postoperative complications. She underwent inpatient rehabilitation and was discharged from the hospital 4 weeks after the operation. At her 6-month follow-up evaluation, she was asymptomatic and doing well.

Discussion

The bilateral recurrence of atrial myxomas is rare. Most cases of recurrence are associated with familial forms of myxomas, especially in Carney complex, a syndrome characterized by cardiac myxomas, endocrine abnormalities, skin pigmentation, and neural



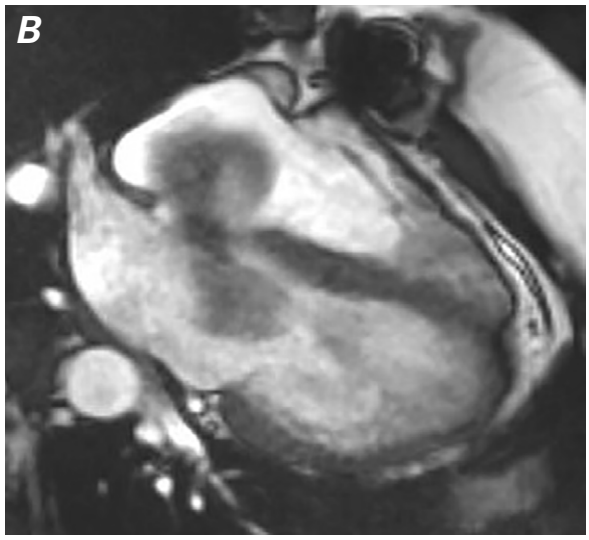
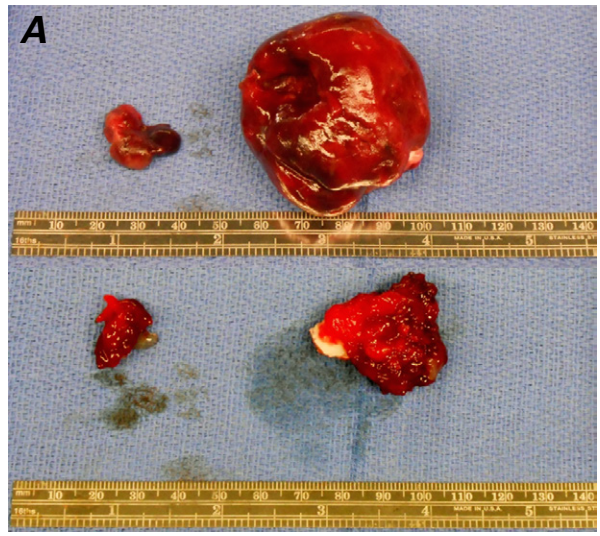


Fig. 1 ***A****) Transthoracic echocardiogram (short-axis view) and* ***B****) cardiac magnetic resonance image (during diastole) show the right (RA) and left atrial (LA) myxomas.*



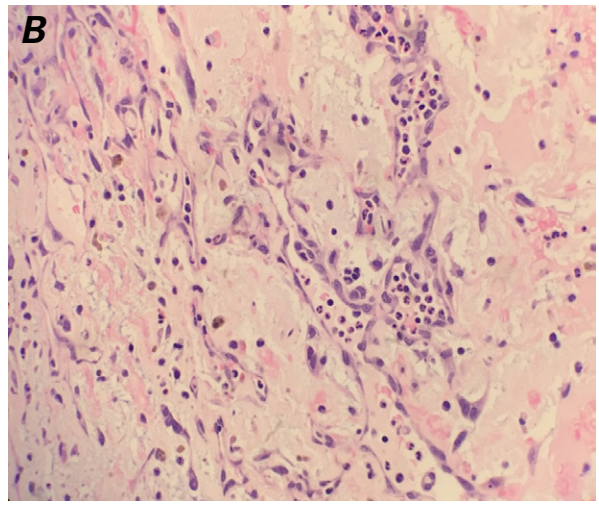


Fig. 2 ***A****) Photograph shows gross specimens of the right atrial (top) and left atrial myxomas.* ***B****) Photomicrograph shows mesenchymal stellate cells in a myxoid stroma (H & E, orig. ×40). Tissue samples were also positive for CD31 and calretinin (not shown).*

disease.[1] Irani and colleagues conducted a literature review and found only 30 documented cases of biatrial myxomas.[2] The frequency of recurrence is about 22% for complex forms and 12% for other familial forms, whereas the frequency of recurrence is 1% to 3% for sporadic myxomas, which seldom recur after removal.[3] Sporadic multifocal recurrences—growths of tumors at multiple sites within one cardiac chamber or in multiple chambers—are even rarer.[4-6]

Multifocal recurrence may result from intracardiac implantations of embolic fragments that break off from the original tumor, either spontaneously or because of damage during surgery.[5,6] Similarly, some myxomas undergo malignant transformation, which leads to spread. Cases have been reported of histologic progression of tumors exhibiting sarcomatous features and of myxomas extending, or even metastasizing, to extracardiac sites.[7,8]

Familial syndromes, such as Carney complex, have substantial potential for multifocal recurrence. Defects at several loci on chromosomes 2, 12, and 17 have been implicated. In particular, researchers studying cardiac myxomas found that the *PRKAR1A* gene, which encodes the regulatory subunit of a protein kinase, was mutated in up to 80% of patients with Carney complex.[9] Of note, investigators have found little to no association between the *PRKAR1A* gene or the aforementioned loci in sporadic myxomas.[10,11] These cases most often involve chromosomal regions 12p1 and 17p1, which suggests that sporadic myxomas may be associated with certain genetic susceptibilities that are distinct from those of familial origin.[10,11]

Some gene-expression data suggest that simultaneous myxomas develop from multiple pretumorous foci that are remnants of embryonic heart development. These data raise the possibility that myxomas derive from primitive, multipotent mesenchymal cells that are phenotypically similar to the primordial cardiac stem cells found in the fetal endocardium.[9,12] If this is the case, islands of these precursor cells may persist, with the potential to proliferate later. As described above, certain genetic predispositions may lead to activation of these scattered vestiges, resulting in multicentric growth in the adult heart.

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