Carney Complex with Right Ventricular Myxoma following Second Excision of Left Atrial Myxoma

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We report a case of Carney complex with massive right ventricular myxoma after two-time excision of a left atrial myxoma. The patient was a 45-year-old woman with pyrexia. She temporarily lost consciousness during examination, and echocardiography and computed tomography (CT) showed a massive tumor in the right ventricle. Loss of consciousness was determined to be caused by intracardiac obstruction of blood flow due to the tumor, and corrective surgery was performed. Pathological findings indicated myxoma with no malignancy. Myxomas are benign, but there is frequent recurrence of tumors associated with Carney complex. Because her myxomas were accompanied by unusual skin pigmentation, she was diagnosed with Carney complex. Carney complex has a high rate of myxoma recurrence, and often runs in families. In all cases, it is necessary to observe the patient’s course closely.

Keywords: Carney complex, myxoma, recurrence

Introduction

Myxomas are the most common type of cardiac tumor. Typically developing in middle-aged women, single myxomas are often found in the left atrium. Recurrence following surgical excision is rare, with an incidence of only 1%–3% of all myxomas, and the postoperative course is generally good. However, some myxomas occur as part of a syndrome known as Carney complex, which have some unusual features. Carney complex, which was first reported in 1985 by Carney, et al., is a syndrome in which myxomas of the heart, skin, and breast are accompanied by skin pigmen-

tary abnormalities as well as endocrine tumors/overactivity and schwan-

omas. In Carney complex, myxomas may occur in any or all cardiac chambers, and recurrence rates are high. Furthermore, other family members are often similarly affected, so accurate diagnosis and careful management of these patients are necessary.

In this report, we review the literature and present a case that was diagnosed as Carney complex based on the clinical characteristics.
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detailed examinations. The patient temporarily lost consciousness during the examinations and was immediately admitted. An echocardiogram showed a massive tumor occupying a large area within the right ventricle. Computed tomography (CT) also showed a massive tumor in the right ventricle (Fig. 1). Because the syncope was thought to have been due to intracardiac obstruction of blood flow caused by the right ventricular tumor, she was referred to our department for surgery. A blood culture was performed to investigate the causes of the pyrexia, and *Staphylococcus aureus* was detected. In addition, findings suggestive of vegetation on the posterior cusp of the mitral valve were observed along with mild mitral regurgitation. Early surgical intervention was planned for removal of the right ventricular tumor accompanied by infective endocarditis. However, while the patient was awaiting surgery, intestinal ischemia and cerebral infarction with transient right arm paresis occurred that was likely due to the infective endocarditis. Thus, surgery was performed 15 days after improvement of her general condition. The electrocardiogram showed normal sinus rhythm.

For the surgery, we began extracorporeal circulation with femoral artery perfusion and femoral vein drainage, and we approached the heart via a median sternotomy (the patient’s third). A tumor was observed through a right atriotomy. The tumor was massive with a smooth surface and gelatinous appearance, and it prolapsed from the right ventricle over the tricuspid valve into the right atrium (Fig. 2). It adhered to the chordae of the anterior leaflet of the tricuspid valve, the anterior papillary muscle and anterior wall of the right ventricle. We excised the tumor as much as possible. We reconstructed the chordae of the anterior leaflet with artificial chordae, which had been excised along with the tumor, and also performed tricuspid valve annuloplasty with a 28-mm Cosgrove-Edwards annuloplasty band (Edwards Lifesciences, Irvine, California, USA). No other tumors were found in either the right atrium or right ventricle. Mitral valve regurgitation was present, which was accompanied by hypertrophy and sclerosis of the posteromedial commissure of the valve leaflet and annulus, where the Kay annuloplasty had been employed in the previous surgery. We repaired the posteromedial commissure. There were neither clear signs of vegetation or neoplasm on the valve leaflet, nor were there any neoplasms in the left atrium. The postoperative pathological examination indicated myxoma with no malignant findings. Her postoperative course was good, and the patient’s condition is currently being monitored through outpatient visits. After the surgery, cancer of the right breast was discovered and excised. The lesion was a papillary carcinoma accompanied by serous fluid.
Discussion

Carney complex, which was reported in 1985 by Carney, et al., is characterized by skin pigmented abnormalities, myxomas, endocrine tumors/overactivity and schwannomas.\(^2\) Research findings have suggested that its onset is related to mutation in the \textit{PRKAR1A} gene.\(^3,4\) The criteria for the clinical diagnosis of Carney complex are two or more major manifestations such as spotty skin pigmentation lesions, myxomas, endocrine tumors or overactivity, schwannoma, or breast ductal adenoma. In addition the diagnosis can be made if there is one of these major manifestations in combination with the supplementary criteria of either an affected first-degree relative or inactivating mutation of the \textit{PRKAR1A} gene.\(^5\) Our patient exhibited a recurrent cardiac myxoma and skin pigmentation lesions on the face and trunk. The diagnosis was Carney complex, because she had two of the major manifestations described above. There were no clear findings of the other major manifestations of endocrine tumors/overactivity or schwannoma. Breast ductal adenomas are considered to be common among Carney complex patients, and this patient had comorbid intracystic papillary carcinoma. The patient had atypical psychosis, and would not give consent for molecular genetic testing. Since her family history was unknown, it is unclear whether she met any of the supplementary criteria for the diagnosis.

According to Edwards, et al., the mean age of onset of Carney complex is 26 years old.\(^6\) It characteristically occurs in young people, with a higher prevalence in females (62%). The site of occurrence is the left atrium in 64% of cases, the right atrium in 44%, the left ventricle in 14% and the right ventricle in 12.5%. In 41% of cases, there are multiple lesions, with 9% occurring in a single chamber and 31% occurring in two or more chambers. The rate of recurrence is 20%; the tumor recurs more than twice in half of the cases with recurrence. In 52% of cases, similar tumors occur in family members; and in cases confirmed to be hereditary, the mode of inheritance is autosomal dominant. The myxoma in our case occurred in the right ventricle of a young female patient, who had two previous occurrences in the left atrium.

Myxomas are generally considered to be benign tumors, but cases of recurrence have been reported.\(^7\) Since the myxomas that occur in association with Carney complex often occur at multiple sites or recur in many cases, this syndrome should be suspected in cases of myxoma recurrence. It is necessary to check carefully for the presence of myxomas in other locations prior to and during surgery. Because there is a possibility that lesions may be present in sites that are difficult to confirm preoperatively with diagnostic imaging, we believe that all of the cardiac chambers should be explored during surgery. In addition, the patient will need to be carefully monitored postoperatively for myxoma recurrence on a long-term basis.

The patient described in this report is at high risk for recurrence of myxoma and will need careful follow-up. The patient also has a twin brother, whose course will also need to be observed.

Conclusion

We treated a patient who developed a right ventricular myxoma 17 years after she had surgical excision of a left atrial myxoma twice. Although myxomas are classified as benign tumors, those that are associated with Carney complex tend to recur or occur at multiple sites. This was a case of a second recurrence in a young patient, and she will require strict observation in the future.

Disclosure Statement

The authors have no conflict of interest to declare.

References