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CASE REPORT

Recurrence of cardiac myxoma in the right atrium with Carney complex following resection of myxomas in both ventricles

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Abstract
We describe a case of Carney complex (CNC), a rare hereditary condition, that resulted in the development of cardiac myxomas. We would like to emphasize that we detected the first myxomas in both ventricles, followed by the second myxoma in the right atrium, although cardiac myxomas often originate in the left atrium. We highlight the heightened risk of recurrence and emphasize the importance of performing regular ultrasonic cardiac echography to preclude such outcomes.

Keywords Carney cpmlex (CNC) · Cardiac myxoma · Recurrence

Introduction

Carney complex (CNC) is a rare hereditary endocrine tumor disease. Approximately 20–40% of CNC patients have a cardiac myxoma. However, tumors in both ventricles of the heart are very rare. We report the case of a cardiac tumor recurring in the right atrium after surgical resection of tumors in both ventricles.

Case report

A 47-year-old woman visited our hospital for a follow-up examination of CNC. Since her childhood, the patient had regularly undergone benign skin tumor resection. She also had also pale, brown, and black lentigines. By a close inspection at the age of 44 years, the patient was found to have hyperepinephry with a left adrenal gland tumor, a thyroid tumor, and cardiac tumors. Echocardiography and cardiac magnetic resonance imaging (MRI) showed both a pedunculated tumor (30 mm in diameter) in the left ventricle and a caulescent tumor (10 mm in diameter) complicated with tricuspid regurgitation in the right ventricle (Fig. 1a). At the time she underwent cardiac surgery for the resection of both tumors. Cardiopulmonary bypass was established between the ascending aorta and the superior and inferior vena cava through median sternotomy. After aortic cross-clamping and cardiac arrest induction using cardioplegia through right atriotomy, the top of the jelly-like tumor was found to be partially stuck in the posterior leaflet of the tricuspid valve. It was mobile, had a narrow stalk and was located in the right ventricle. The bottom of the stem tightly adhered to the myocardium of the right ventricle involving the posterior leaflet of the tricuspid valve, very close to the anterior leaflet. The tumor in the right ventricle (3 cm in diameter) was resected en-bloc with both chordae of the posterior leaflet and part of the posterior leaflet itself (Fig. 1b). Then, via a trans-atrial septal approach, the tumor in the left ventricle was inspected through the mitral valve. The jelly-like tumor was 1 cm in diameter and attached to one of the primary chorda of the posterior leaflet of the mitral valve; the tumor did not touch the posterior medial papillary muscle of the left myocardium. As the tumor in the left ventricle was resected en-bloc, the area was thoroughly investigated to confirm that no other tumors were present in the left ventricle, including the mitral valve and its apparatus. The saline test was performed to confirm no mitral regurgitation. After closing the septal wall and repairing the right atrium, the cardiac beat restarted following removal of the aortic cross clamp. However, transesophageal cardiac echography revealed severe tricuspid regurgitation after weaning the patient off cardiopulmonary bypass. Cardiopulmonary bypass was restarted, and the tricuspid valve was repaired using a modified Kay's method, involving annulorrhaphy of

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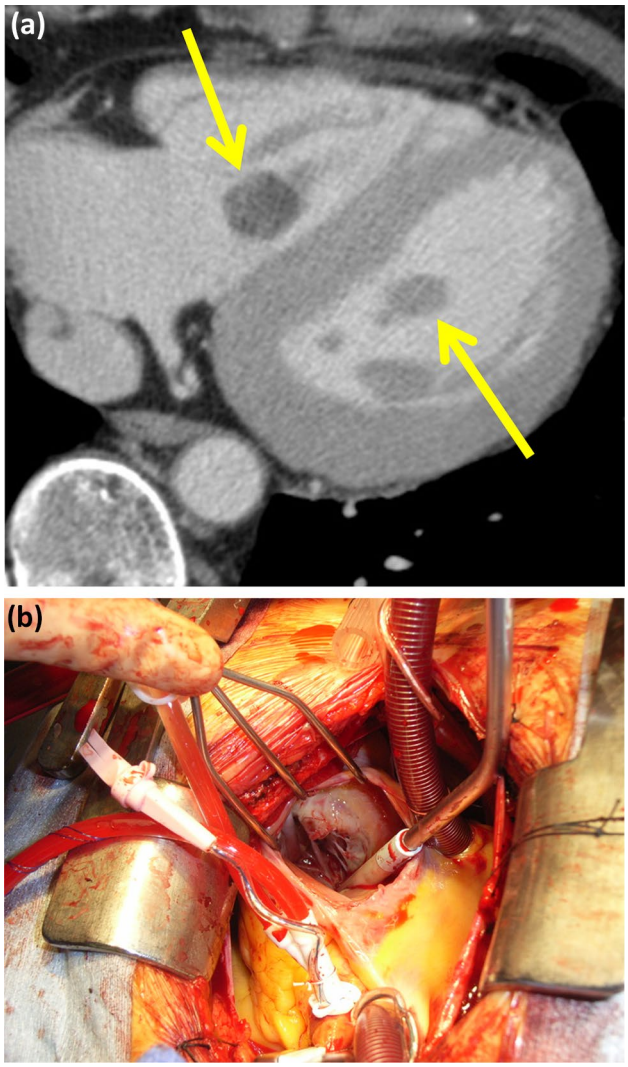

Fig. 1 Enhanced computed tomography revealed each tumor originating either left ventricle or right ventricle (**a**). Operative findings showed jelly-like tumor, which was originating in the right ventricle, adhering tricuspid posterior leaflet (**b**)

the posterior segment. Then, the second attempt to wean the patient off cardiopulmonary bypass was uneventful, resulting in trivial tricuspid insufficiency. Thereafter, myxomas with both tumors were definitively diagnosed by a pathology examination. After the left adrenal gland tumorectomy, the patient was stable for two and a half years. At 3-year postcardiotomy, her echocardiography incidentally showed recurrence of the cardiac tumor, which was in the right atrium; enhanced CT was used to thoroughly examine whether the tumor was only in the right atrium (Fig. 2a, b). Because severe tricuspid regurgitation was detected (data not shown), with resection of the tumor, we planned tricuspid annulo-plasty. As a result of consultation to endocrinologists, there was no abnormality in the endocrinologic blood test of the patient (data not shown). After re-sternotomy, we established cardiopulmonary bypass between the ascending aorta and the superior vena cava via the right femoral vein. The tumor was on the eustachian valve very close to the inferior vena cava (Fig. 2c). We resected the tumor and the attached myocardial tissue. However, the tumor in the right atrium was not associated with tricuspid regurgitation, and dilatation of the tricuspid valve after the Kay repair was noted. Then, we performed tricuspid annulo-plasty using a prosthetic ring and directly closed the right atrial incision. Her operative course was uneventful. The pathological findings were cardiac myxoma. The patient has remained free of cardiac tumor for at least 2 years.

Discussion

CNC itself is a rare genetic disease characterized by skin pigmentation abnormalities, endocrine tumors, schwannomas, and myxomas [1]. It was described for the first time by Carney [2]. The heredity pattern is autosomal dominant, and while PRKAR1A to contain the causative gene, the gene itself remains unknown [1, 3, 4]. Approximately 20–40% of CNC patients have a cardiac myxoma [1, 5]. Among all cardiac tumors, CNC-related tumors account for 7% [6]. The most common site of origin is the left atrium, followed by the right atrium and ventricle; in some cases, tumors originate in both chambers [5, 7]. Cardiac tumors cause 50% of deaths in CNC. These tumors can cause sudden death due to the effects of tumor embolism, such as stroke or acute valve blockage [7, 8]. Patients are most commonly in their twenties when they are diagnosed, and it seems they can lead a normal life if their tumors are treated; however, some patients die young [1, 8]. In CNC, the rate of recurrence is as high as 22%, whereas the recurrence rate in isolated cases is approximately 1–3% [9]. Although a cardiac surgeon is often the first to detect these tumors, patients management needs to involve genetic and endocrine experts. We provide evidence in support of relative genetic testing to preclude the possibility of developing CNC and the associated myriad of tumors. Moreover, endocrinologists must be consulted in such cases for disturbances in the endocrine system.

Conclusion

We reported a rare case of cardiac myxoma recurrence outside the left atrium, in associated with CNC. CNC should be included in the differential diagnosis when the surgical removal of a cardiac myxoma is required.

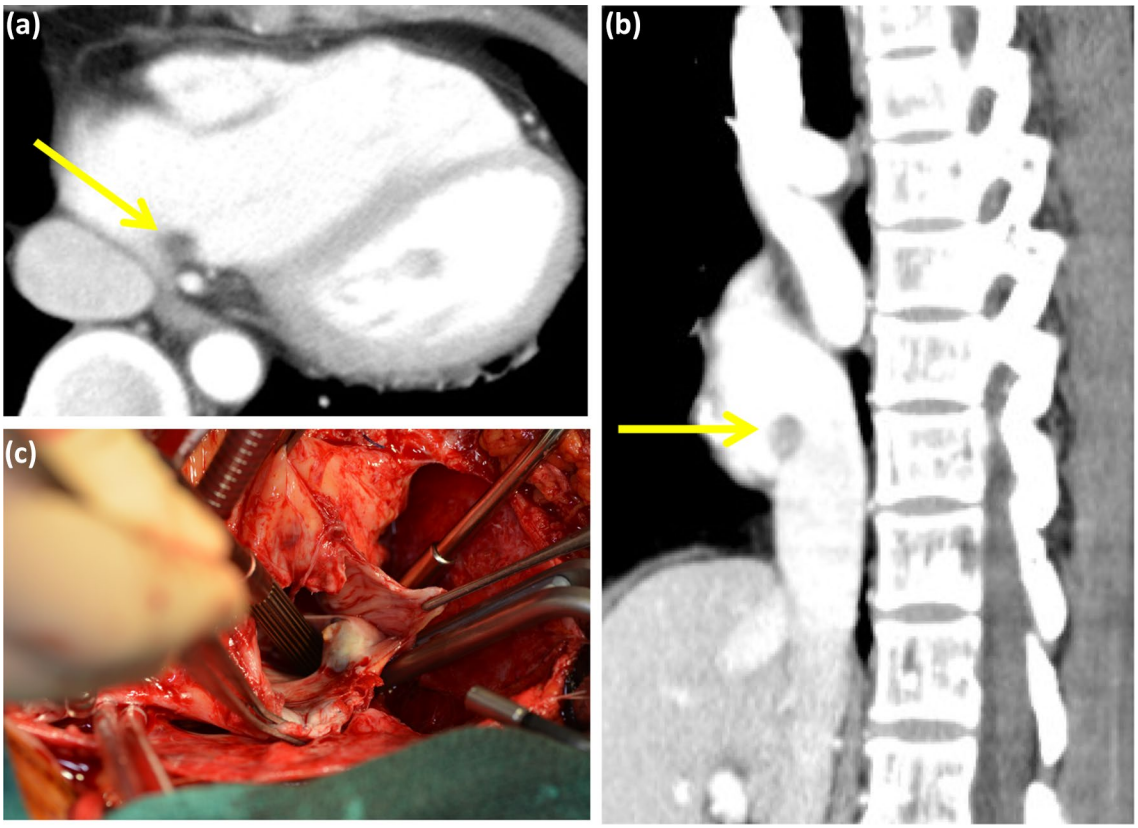

Fig. 2 Axial and sagittal view of enhanced computed tomography revealed that there was a tumor near the inlet of inferior vena cava in the right atrium. There were no recurrence of tumor in other cardiac chambers (data were not shown) (**a, b**). Operative findings showed a smooth surface tumor, which was originating in the right atrium, near the Eustachian valve, behind clamp of IVC (**c**)

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Compliance with ethical standards

Conflict of interest The authors declare no conflicts of interest.

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