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Case Report

Carney Complex with Biatrial Cardiac Myxoma

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Cardiac myxomas make up approximately 50% of all benign cardiac tumors and represented 86% of all surgically treated cardiac tumors. Most of them originated from the left atrium, in some cases from both of atria. We report a case of male patient with biatrial myxomas and other extra-cardiac involvement: hypophyseal adenoma, enlargement of thyroid gland, tubular adenoma polyp of colon and bilateral large cell calcifying Sertoli cell tumor (LCCSCT) of testis. These findings led to the diagnosis of Carney's complex, which is a syndrome with multiple neoplasias, cardiac myxomas, lentigines, and endocrine abnormalities. A genetic test confirm this diagnosis.

Keywords: cardiac myxomas, Carney's complex, endocrine abnormalities

Introduction

More than 75% of primary cardiac tumors are benign, and most of them are myxomas.[1)] The incidence of myxoma is 0.5 per million population per year.[2)] Up to 80% of them are localized in the left atrium, 7%–20% are found in the right atrium, the rests of up to 10% each are in the right ventricle (2.5%–6%), or in the left ventricle (8%), two or more location (2.5%), and in rare cases, in both atria of the same patient (<2.5%).[3)] When biatrial, the myxomas can arise from mirror-image regions on both side of the septum. The symptoms have varied greatly, depending on the size and the localization of this tumor and may include shortness of breath, dizziness, palpitations, symptoms due to cerebral embolization or pulmonary hypertension. Presence of biatrial myxoma should raise suspicion of Carney's complex. It is characterized by familial recurrent myxomas, pigmented skin lesions and endocrine neoplasms.

Case Report

A 35-year-old white man was admitted for resection of the biatrial tumors. His medical history included partial resection of hypophysis adenoma in 2010, subsequently postoperative hypopituitarism, hypocorticism, peripherial hypogonadism as the result of bilateral large cell calcifying Sertoli cell tumor of testis, partial strumectomy due to struma nodosa, polypectomy of a colonic tubular adenoma. The palpitation was the only symptom of myxomas. Clinical examination and laboratory results were normal. An electrocardiogram revealed sinus rhythm. Echocardiogram showed bilateral atrial tumors arising from the free walls of atria protruding into the left and right ventricles during diastole (**Fig. 1**). Surgical excision of the myxomas was performed under extracorporeal circulation and moderate hypotermia, median sternotomy and a right atriotomy was used. In the right atrium was a fragile tumor 8 × 8 × 5 cm, arised from the fossa ovalis and spreading to the inferior vena cava ostium. The tumor was removed and through the trans-septal approach

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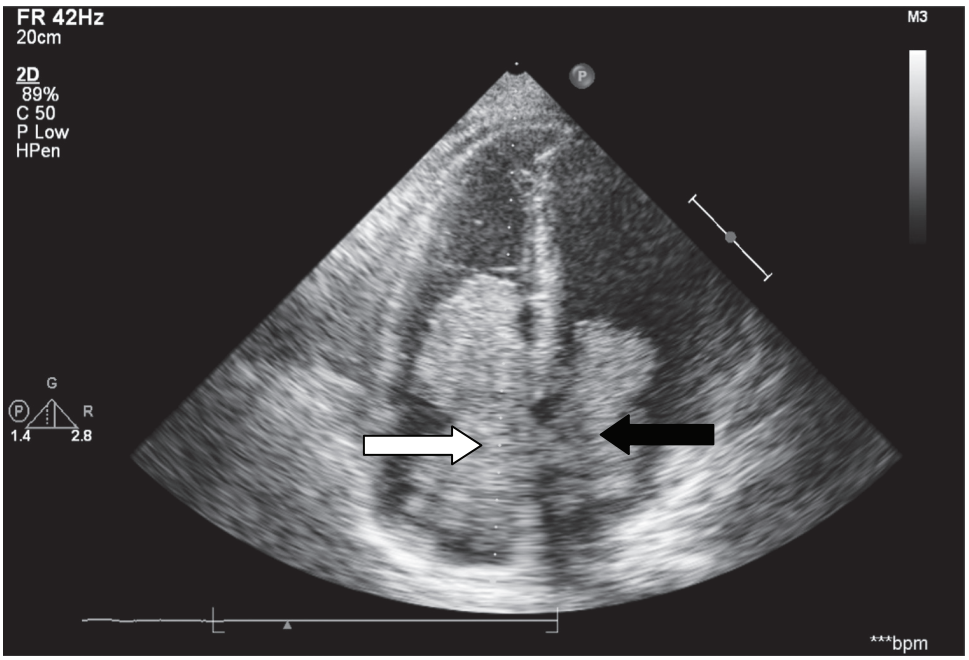

Fig. 1 Echocardiogram showing bilateral atrial tumors (white arrow: right atrial tumor, black arrow: left atrial tumor) with protrusion into the left and right ventricle.

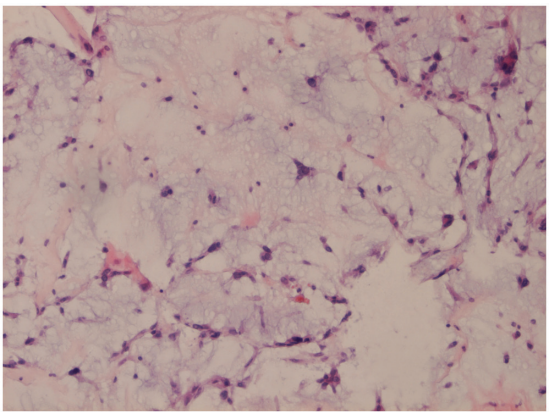

Fig. 2 Cardiac myxoma with low cellularity, tenuous myxoid stroma in the background, dispersion stellate, and spindle mesenchymal myxoma cells with eosinophilic cytoplasm, without significant nuclear atypia, sporadic vascular structures, and the addition of non-specific inflammatory cellulization.

was revealed and resected the left atrial myxoma. This tumor was of identic consistence, 6 × 5 × 3 cm, attached to the free wall of the atrium by a small stalk 3 × 1 cm. The diagnosis of bilateral atrial myxomas was confirmed by histologic studies (**Fig. 2**). We assumed association between the endocrine abnormalities—hypophyseal adenoma, struma nodosa and benign tumors (bilateral LCCSCT of testis and biatrial tumor), and therefore we signed this patient with Carney complex. Carney complex is an autosomal dominant condition most commonly caused by mutations in the PRKAR1A gene on chromosome 17q23-q24. In our patient mutation c.491-492delTG/p.V164Dfs*5 in exon 4 of PRKAR1A gene was identified and the diagnosis of Carney complex was confirmed (**Fig. 3**). The patient was discharged in good condition.

Discussion

Although primary tumors of the heart are rare, the myxoma is the most frequent benign primary heart tumor as it accounts for 0.3% of open-heart surgery.[4] Cardiac myxomas usually appear as a sporadic isolated condition in the left atrium of middle-aged women with no other coincidental pathology. A small percentage of primary cardiac tumors have familial penetrance. Recurrence is more frequent in patients with the family history of myxoma, and familial myxomas frequently appear et early ages, with atypical and multicentric location.[5] In 1985, Carney and others described in young people a special complex group of cardiac myxomas associated to a distinctive complex pathology, giving identity to the

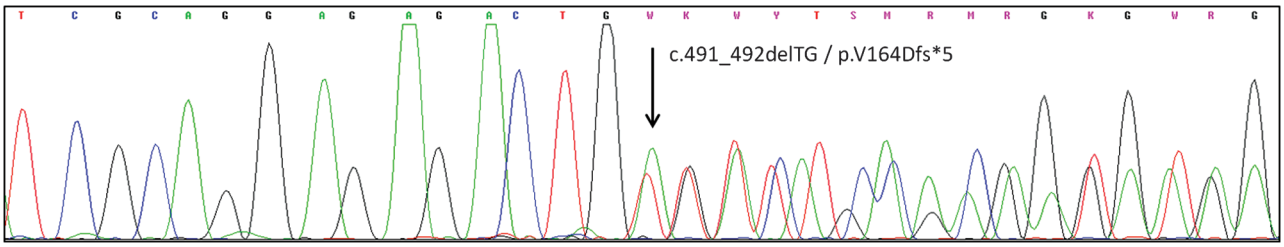

Fig. 3 The Sequence of part exon 5 of PRKAR1A gene. Arrow shows frameshift mutation – deletion of two nucleotides T and G at position 491 and 492 of CDS resulting in frameshift and premature STOP codon.

"Syndrome Myxoma" or "Carney's Complex." Carney's complex (CNC) is a multiple neoplasia syndrome featuring cardiac, endocrine, cutaneous, and neural tumors, as well as a variety of pigmented lesions of the skin and mucosae. Patients with CNC are usually younger, with a mean age of 26 years and female predominance (62%) There is a marked familial trend (52%), a high incidence of recurrence (20%).[6] The most frequent extra-cardiac involvements are pigmented skin lesions, cutaneous myxomas, adrenal cortical disease, myxoid mammary fibroadenoma, pituitary adenoma, melanotic schwannomas, thyroid disease and male patients with testes tumors. The diagnosis is made when two or more of these criteria are present. The myxom in CNC is usually in left atrium, biatrial myxomas are rare. The four chambers of the heart should be examined at surgery for typical myxoma locations, right atriotomy and combined superior-transseptal approach improve exposure of the cavities. In our patient, the tumor size and attachment were identified by echocardiogram, and a safe and efficient transseptal incision was achieved. Myxoma is known to recur in 30%–70% of patients with familial myxomas. Close follow-up of recurrent cardiac myxoma or other tumors is recommended, as is careful screening of family members. Careful screening of the first degree family members should be conducted, and closed short and long term follow up controls are important. There was no recurrence one year after tumor resection in our case. We examined the first degree family members, they were without mentioned clinical features and myxomas of the heart and also the genetic tests were negative.

Conclusion

In conclusion, the presence of multiple myxomas and endocrine abnormalities should be an indication to investigate for the Carney complex.

Disclosure Statement

There are no conflicts of interest, sources of financial support, corporate involvement, patent holdings, etc. for each author.

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