Case report - Cardiac general

Right atrial lipoma in patient with Cowden syndrome

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Abstract

Primary cardiac tumors are rarest form of cancer and the lipoma represent about 8% of these tumors. Cowden disease is a rare autosomal dominant disorder, associated to a germline mutation of the PTEN gene, characterized by multiple hamartomas and an increased risk of breast, thyroid and endometrial carcinomas. For the first time, we describe a right atrial lipoma in a patient affected by Cowden syndrome. The patient suffered of some episodes of atrial flutter. The echocardiogram showed a cardiac mass, suggestive of lipoma with cardiac magnetic resonance images. Right atrial mass was completely resected and the histological examination confirmed the diagnosis of lipoma. The patient was discharged from hospital without any complications.

Keywords: Cardiac lipoma; Cowden syndrome; Cardiac tumors

1. Introduction

Primary cardiac tumors are very rare, ranging from 0.001 to 0.28% of all tumors, and the lipoma is responsible for about 8% of them [1]. Cowden disease, one of the several hamartoma syndromes, is characterized by hyperplastic lesions and hamartomas distributed on the whole body [2]. We describe the first case of right atrial lipoma as a clinical manifestation of Cowden syndrome.

2. Case report

A 62-year-old male with an history of hypertension, cerebral transient ischemic attack and affected by Cowden syndrome with gastrointestinal hamartomas, was referred to us for a cardiac mass into the right atrium. The presence of multiple gastrointestinal hamartomas is not needed for the diagnosis of Cowden syndrome, but it is one of the clinical manifestation of this disease. In our case, the diagnosis of Cowden disease was based on Consortium criteria and afterwards it was confirmed by the detection of the mutation in PTEN gene.

After the recent onset of an episode of atrial flutter, the patient underwent a transthoracic echocardiogram that showed an atrial mass, arising from the atrial septum and taking up more than the half of the right atrium.

The cardiac magnetic resonance images (MRIs) confirmed the relationship between the tumor and the atrial septum that was widely infiltrated.

Moreover, the signal behavior of the mass, identical to fatty tissue, was suggestive of a cardiac lipoma, associated with a cystic component (Fig. 1a and b).

A median sternotomy was performed. Cardiopulmonary bypass was initiated with distal ascending aorta cannulation and bicaval venous cannulation. The tumor appeared to arise from the atrial septum aneurysm, with a large base of plant. The mass and the part of atrial septum infiltrated were completely removed. We preferred to close the surgical defect with a direct running suture as the size of the septal aneurysm allowed for it. The histological examination confirmed the diagnosis of cardiac lipoma (Fig. 2a, b).

The patients was discharged from hospital without any problems after one week.

3. Discussion

Primary cardiac tumors are very rare and about 75% of them are benign [2]. These tumors are often asymptomatic until they grow enough to cause symptoms. Cardiac lipomas are 50 times less common than myxoma and they grow very slowly. They usually arise from the epicardial fat tissue, growing into the pericardial sac. The most frequent intra-cardiac location is the right atrium, where they can originate either from the atrial septum or the atrial roof. The symptoms are usually caused by the overgrowth and they are dyspnea, embolism, atrial and ventricular arrhythmias [3].

Cowden syndrome is a rare autosomal dominant disorder, associated to a germline mutation of the PTEN gene, characterized by multiple hamartomas and an increased risk of breast, thyroid and endometrial carcinomas. The diagnosis of Cowden disease was based on the detection of the mutation in PTEN gene.

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Fig. 1. A cardiac magnetic resonance images (MRI) show a huge right atrial mass with a cystic components, arising from atrial septum (a and b). The signal behavior is suggestive for cardiac lipoma.

Fig. 2. (a) Histological examination confirm the diagnosis of cardiac lipoma in Cowden disease. (b) High-power microscopic view.

risk of breast, thyroid and endometrial carcinomas. The subcutaneous lipomas and cutaneous hemangiomas often appear [4–6].

We have searched in the literature for the cardiac involvement in Cowden disease without finding any citations.

We want to underline certain features of the management of our case. First, the role of cardiac magnetic resonance imaging (MRI) to identify the nature of the cardiac tumor and its relationship with the near structures, allowing to plan the best surgical approach.

Second, heart can be involved in Cowden syndrome, that caused the growth of cardiac tumor, such as lipomas, that often remain asymptomatic for long time. Therefore, echocardiography is an important tool to discover cardiac involvement of Cowden disease before growing enough to become symptomatic.

We conclude that Cowden syndrome can cause right atrial lipoma and in this case the mainstay of the treatment remains the complete surgical resection, even if these tumors can grow again.

References

After reading the interesting case report on right atrial lipoma in a patient with Cowden syndrome [1] we would like to make a few remarks. Although echocardiography is considered to be the most important diagnostic technique in such cases, magnetic resonance imaging can provide detailed characteristics and best visualization of the tumours. We are also aware that plain chest radiography findings are sometimes suggestive of primary cardiac tumours but not pathognomonic. Where there any suggestive findings of the tumour on a plain chest X-ray film, for example, a notable calcification or a widened mediastinum?

We also assume that the authors performed a complete excision of the tumour reaching negative margins. In regard to the created defect, do they routinely use a direct running suture for the repair or do they consider a patch also?

Finally, unlike cutaneous lipomas, cardiac lipomas present a rather confused genetic and molecular background. Vaughan et al. [2] reported an unusual translocation between chromosomes 2 and 19 in a patient with a large invasive cardiac lipoma. Besides the mutation in PTEN gene which was detected by the authors, were any cytogenetic studies indicative of the aforementioned perturbation?

References
