Recurrence of Carney complex atrial myxoma causing Lung nodules

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Abstract

Background Myxomas are the most common form of benign primary cardiac tumor, most of them are located in the left atrium. When they are located in the right atrium, It may occur with pulmonary artery embolism. **Methods and Results** A 72-year-old woman had endured more than 5 months with chest tightness and difficulty breathing. Based on various clinical findings and data, we diagnosed this subject as Cushing's syndrome. Emergency bedside ultrasound showed right atrial tumor, which was confirmed by coronary computed tomography angiography (CCTA).The patient underwent surgical resection of the right atrial and pulmonary artery tumors.In the two subsequent short-term reviews (one month and half a year after surgery), we found same new myxoma in the atrial septum and pulmonary nodules. **Discussion** Tumor recurrence is uncommon, although not unknown, after complete excision. 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. **Conclusion** They face a mottled skin pigmentation, and gorgeous features of Cushing's syndrome. Myxomas recurrence have been reported in numerous reports. In contrast with other reported cases, a specific characteristic of the present case was the short-term recurrence of Cardiac myxoma and newly grown nodules in the lungs. Patients received our recommendations for lifetime anticoagulation therapy and long-term follow-up observations.

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Discussion Tumor recurrence is uncommon, although not unknown, after complete excision. 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.

Conclusion They face a mottled skin pigmentation, and gorgeous features of Cushing's syndrome. Myxomas recurrence have been reported in numerous reports. In contrast with other reported cases, a specific characteristic of the present case was the short-term recurrence of Cardiac myxoma and newly grown nodules in the lungs. Patients received our recommendations for lifetime anticoagulation therapy and long-term follow-up observations.

KEYWORDS

Carney cpmlex (CNC), Cardiac myxoma, Recurrence

A 72-year-old woman had endured more than 5 months with chest tightness and difficulty breathing. Her previous medical history was that she had interventional occlusion surgery for atrial septal defects 8 years ago. On physical examination we found that she had a full moon face and central obesity signs, spotty skin pigmentation lesions,while urinary 17-hydroxycorticosteroid measurements were significantly elevated. Based on various clinical findings and data, we diagnosed this subject as Cushing's syndrome.

Emergency bedside ultrasound showed right atrial tumor, which was confirmed by coronary computed tomography angiography (CCTA). CCTA showed uneven low-density clumps attached to the edge of the blocker, diastolic phase protruding into the right chamber, and systolic phase completely retracting into the right atrium (Panels A, white arrow, Supplementary material online, Videos S1 and S2). Pulmonary artery multiple filling defect, was attached to the wall, free and completely blocked the lumen, embolization index 70% (Panels C, white arrow).

The patient underwent surgical resection of the right atrial and pulmonary artery tumors, partial removal of atrial septal occluder and atrial septal repair. To ensure complete removal of the tumor, we resected with a portion of tumor-free atrial septum, followed by primary reconstruction of the septum. During the operation, we found that the right chamber was gray, jelly sample, brittle tissue, attached to the atrial septum and the edge of the blocked umbrella. Histopathological examination showed stellate mesenchymal cells in a myxoid stroma (Panels B, Stellate-shaped myxoma cells).

For CCTA review before discharge, the right atrial surface of the atrial septum had less low-density shadow and 5mm thickness, (Panels E, white arrow, Videos S3), No abnormal density was observed in each cardiac cavity. However, the CCTA was reviewed in 6 months after surgery, the low density shadow significantly increased, the thickness was 11mm (Panels F, white arrow, Videos S4). Pulmonary embolism improved, with an embolization index of 50% (Panels G, white arrow). New nodules were found in both lungs, short diameter of 3mm-20mm(Panels H, white arrow,Contrast the preoperative CCTA,Panels D), lung dissemination should be suspected. The patient gave consent for her anonymous clinical data to be published in this report.

Discussion:

Tumor recurrence is uncommon, although not unknown, after complete excision. Recurrence following surgical excision is rare, with an incidence of only 1%–3% of all myxomas, and the postoperative course is generally good [1]. However, some myxomas occur as part of a syndrome known as Carney complex, which have some unusual features. According to Wilbring, et al., elegantly report a 60-year-old female patient with Carney complex and history of previous 4 cardiac operations for recurrent myxoma [2].

Carney complex (CNC) is a rare autosomal dominant syndrome, characterized by mucocutaneous pigmentation, cardiac, cutaneous myxomas and endocrine overactivity. 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 PRKAR1A gene [3]. They face a mottled skin pigmentation, and gorgeous features of Cushing's syndrome. Myxomas recurrence have been reported in

numerous reports[4,5,6]. In contrast with other reported cases, a specific characteristic of the present case was the short-term recurrence of Cardiac myxoma and newly grown nodules in the lungs. Patients received our recommendations for lifetime anticoagulation therapy and long-term follow-up observations.

Figure 1. Computed tomography angiography(CTA) imaging and pathological finding.

CTA revealed uneven low-density clumps attach to the edge of the blocker(A). The pathological finding revealed stellate mesenchymal cells in a myxoid stroma(B). Computed tomography pulmonary arteriography(CTPA) demonstrated massive pulmonary embolism before surgery (C).Chest CT showed no lung nodules before surgery(D). One month after surgery, the CTA showed a low-density filling defect in the right atrial septum (E). Six months after surgery, the CTA showed a large number of low-density filling defect in the right atrial septum (F). Six months after surgery, the CTPA demonstrated pulmonary embolism was improved (G). Six months after surgery, Chest CT showed multiple new round lung nodules in the lungs(F).

Supplementary material online, Videos S1

CTA revealed uneven low-density clumps attach to the edge of the blocker,Cross the tricuspid valve into the right ventricle during the diastolic phase and return to the right atrium during the systolic phase.

Supplementary material online, Videos S2

CTPA confirmed massive pulmonary embolism in the pulmonary arteries.

Supplementary material online, Videos S3

One month after the operation, CT of the mediastinal window showed a new growth low-density filling defect on the right side of the atrial septum, and the CT of the lung window showed no pulmonary nodules in the lungs.

Supplementary material online, Videos S4

Half a year after the operation, ct of the mediastinal window showed a new growth low-density filling defect on the right side of the atrial septum, which was significantly larger than before, and the lung window CT showed multiple new round lung nodules in the lungs.

Reference:

1.McCarthy PM, Piehler JM, Schaff HV, et al. The significance of multiple, recurrent, and “complex” cardiac myxomas. J Thorac Cardiovasc Surg 1986; 91: 389-96.

2.Wilbring M,Wiedemann S,Kappert U,et al.A complicated case of Carney complex: fifth reoperative cardiac surgery for resection of recurrent cardiac myxoma.[J].J Thorac Cardiovasc Surg,2013,4:e22-4.

3. Stratakis CA, Kirschner LS, Carney JA. Clinical and molecular features of the Carney complex: diagnostic criteria and recommendations for patient evaluation. J Clin Endocrinol Metab 2001; 86: 4041-6.

4. Ando T,Goto H,Date K,et al.Recurrence of cardiac myxoma in the right atrium with Carney complex following resection of myxomas in both ventricles.[J].Gen Thorac Cardiovasc Surg,2019,10:891-893.

5. Wei K,Guo HW,Fan SY,et al.Clinical features and surgical results of cardiac myxoma in Carney complex.[J].J Card Surg,2019,1:14-19.

6.Tamura Y,Seki T.Carney complex with right ventricular myxoma following second excision of left atrial myxoma.[J].Ann Thorac Cardiovasc Surg,2014,:882-4.