Case Report

A rare case of renal angiomyolipoma involved both inferior vena cava and pulmonary arteries

Jun Gu[1*], Ling Zeng[2*], Hao Zeng[3], Chaoyi Qin[1], Zhong Wu[1]

[1]Department of Cardiovascular Surgery, [2]Department of Intensive Care Unit, [3]Department of Urinary, West China Hospital, Sichuan University, Chengdu 610041, China

*These authors contributed equally to this work.

Correspondence to: Zhong Wu. Department of Cardiovascular Surgery, West China Hospital, Sichuan University, Lane outside the southern No.37, Chengdu 610041, China. Email: wuzhong71@scu.edu.cn.

Abstract: Angiomyolipoma is a benign mesenchymal tumour, which rarely involve inferior vena cava (IVC) and pulmonary arteries. Here we reported a 41-year-old man, who presented symptoms of chronic pulmonary embolism. After multiple images evaluation, a large tumour with the same properties as the pulmonary embolism was found in the right kidney. One-staged surgery to remove both pulmonary embolism and right kidney was performed. Histological study showed the tumour and embolism as angiomyolipoma. The present study reported a rare case with renal angiomyolipoma and both IVC and pulmonary arteries involvements, which provided a good example for differential diagnosis in the emergency department and treating a rare type of chronic pulmonary embolism after renal angiomyolipoma.

Keywords: Angiomyolipoma; renal; inferior vena cava (IVC); pulmonary artery

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Introduction

Angiomyolipoma, first described in 1900 by Grawitz, is a benign mesenchymal tumour and consists of adipose tissue, spindle and epithelioid smooth muscle cells. It is featured by extensive proliferation of perivascular epithelioid cells (1). It is reported that around 0.1–0.3% prevalence in general population and kidney is involved most commonly. However, it is really rare to find that renal angiomyolipoma invaded into cardiovascular system, especially the pulmonary arteries (2). Most cases of pulmonary embolism happened during the nephrectomy and need emergent pulmonary embolism removal (3,4). Here we presented an interesting case that renal angiomyolipoma and chronic pulmonary embolism were removed in the one-staged surgery.

Case presentation

A 41-year-old man was admitted into our department of emergency due to recurrent cough and activity chest tightness. Medical history and physical examination failed to identify any positive signs. Contrast-enhanced chest computed tomography (CT) and CT angiography showed a tumour with adipose density in the left pulmonary trunk, which partially protruded into the right pulmonary trunk, resulting in the narrowness of left inferior pulmonary artery and its branches. To further understand the tumour, cardiac magnetic resonance imaging was applied, showing a tumour with short T1 and long T2 signal in the left pulmonary trunk, suggesting it as adipose origin (*Figure 1A*). In addition, positron emission tomography-CT was performed, finding a 75×59 mm^2 tumour with adipose density in the right kidney, considering it as an angiomyolipoma, which infiltrated into inferior vena cava (IVC) through right renal vein. After consulting with the medical team of urological surgery, we decided to apply surgical intervention to remove the right kidney and the tumour in the left pulmonary trunk at the same time.

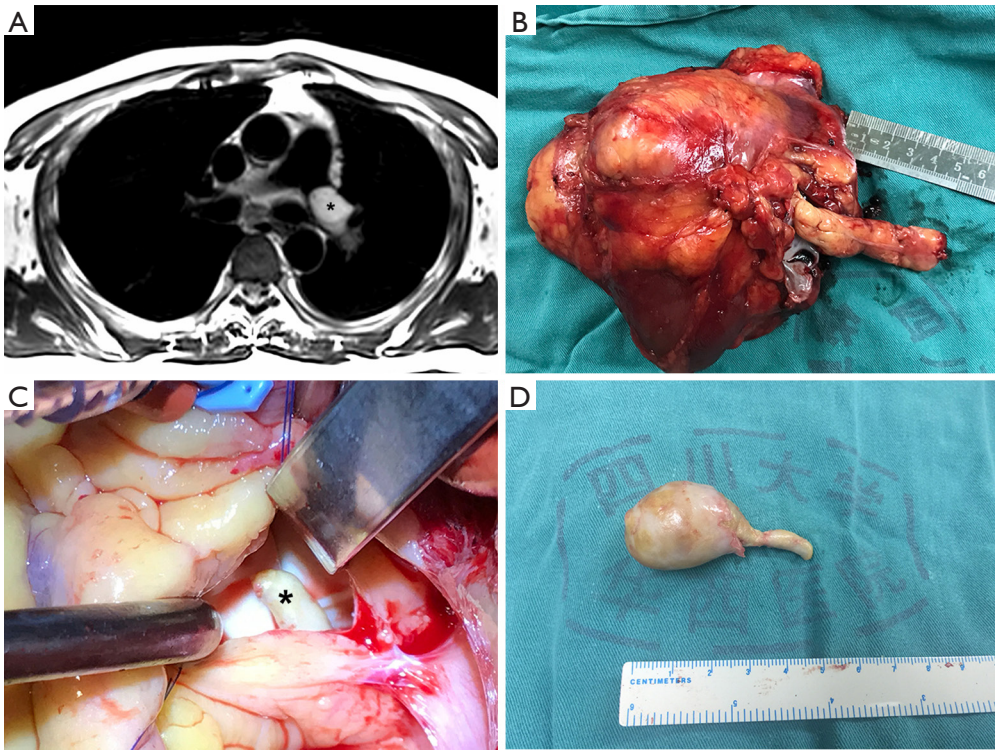

Figure 1 Multiple images of renal angiomyolipoma involved both inferior vena and pulmonary arteries. (A) Cardiac MRI showed a tumour (indicated by the star) with short T1 and long T2 signal in the left pulmonary trunk, suggesting it as adipose origin; (B) gross anatomy during operation showed extensive adipose tissue in the middle of the kidney; (C,D) pedunculated tumour embolus (indicated by the star) in the left pulmonary trunk was removed during operation.

IVC, right renal vein and artery were exposed step by step under general anesthesia. After ligation of right renal artery and clamping of IVC and left renal vein, the entrance of right renal vein into the IVC was cut and tumour embolus was removed carefully. After closing the incision, IVC and left renal vein were reopened. Standard nephrectomy was performed and gross anatomy showed extensive adipose tissue in the middle of the kidney (*Figure 1B*). After closing right abdominal incision, cardiopulmonary bypass was performed. Under deep hypothermic circulatory arrest, left pulmonary trunk was opened and the pedunculated tumour embolus was removed (*Figure 1C,D*). The patient was routinely sent back to the cardiac intensive care unit and transferred into our ward on post-operative day 3. Pathological study showed the mature adipocytes, thick-walled blood vessels and smooth muscle cells, which identified the tumour as angiomyolipoma. The patient was fully recovered and discharged on post-operative day 8.

Discussion

Angiomyolipoma is commonly sporadic and around 20% is associated with tuberous sclerosis complex and lymphangioleiomyomatosis (2). Renal angiomyolipoma can be divided into two histological categories, classic and epithelioid. And most patients with sporadic renal angiomyolipoma are classic, which have better prognosis than epithelioid ones. The epithelioid type presents malignant behaviours, such as invading into the renal vein, IVC, even the heart (5,6).

Pulmonary involvement is a rare presentation in renal angiomyolipoma patients, even in some malignant renal tumours (4). Most patients with renal angiomyolipoma are asymptomatic and usually found by radiological or sonographic images (7). However, patients with renal angiomyolipoma become symptomatic if the involvements of renal vein, IVC or intra-cardiac extension and pulmonary embolism occur (1,8,9). In our case, the patient only suffered chronic pulmonary

embolism, admitted into the emergency department and presenting recurrent cough and activity chest tightness. And laboratory data of the patient showed no kidney dysfunction although the right kidney has no function.

Treatment of renal angiomyolipoma alone is still controversial. Given the compression to the adjacent tissue, the tumours larger than 4 cm are recommended to surgical removal (10). By contrast, with exist of renal vein or IVC thrombus, surgical intervention is indicated regardless of symptoms (8). For patients with life-threatening pulmonary embolism, we believe that the preferred procedure is urgent surgery. And the multi-disciplinary team is necessary for make the surgical plan. In our case, after consulting with medical team of urological surgery, surgical plan of one-staged removing both pulmonary embolism and right kidney was made together (11). And two surgeries will significant increased surgical risks and the residual tumor might increase due to the stimulation of the surgery. And it is reported that concomitant operation had decreased ventilation time and hospital stay (4).

Evaluation by multiple images is of importance as well. Activity chest tightness is the common symptom in the emergency department, which need differential diagnosis carefully. In this case, given the history of chronic recurrent cough, chronic pulmonary embolism is suspected. Pulmonary embolism with malignant cancer is commonly associated with a hypercoagulable state, which is thrombotic (12). Therefore, it is essential to tell the properties of pulmonary embolism before surgical intervention. Meanwhile, in order to locate the origin of pulmonary embolism and find other involvements, full body screen should be performed. As for angiomyolipoma, biopsy is not needed because of typical imaging findings (9). In our experience, MRI and CT are performed to fully evaluate the pulmonary embolism, find IVC involvement, and locate the primary renal angiomyolipoma.

In the present case report, we reported a rare case of renal angiomyolipoma with chronic pulmonary embolism. After multiple images evaluation, renal angiomyolipoma-originated embolism was confirmed, which helped to decide remove the pulmonary embolism and involved kidney at the same time. Our experience provided a good example for differential diagnosis in the emergency department and treating a rare type of chronic pulmonary embolism.

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None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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