

RAPIDLY GROWING RIGHT VENTRICULAR OUTFLOW TRACT MASS IN PATIENT WITH SARCOMATOID RENAL CELL CARCINOMA

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Cardiac metastasis from renal cell carcinoma (RCC) without inferior vena cava (IVC) involvements is extremely rare with few reported cases. Sarcomatoid RCC with rhabdoid feature is a rare pathologic type of RCC having aggressive behavior due to great metastatic potential. Here, we report a case of rapidly growing cardiac metastasis of RCC which brought on right ventricular outflow tract (RVOT) obstruction without IVC and right atrial involvement in a 61-year-old woman. Cardiac arrest occurred during radical nephrectomy and echocardiography revealed mass nearly obstructing the RVOT which was not recognized by preoperative echocardiography 1 month ago. Postoperative immunohistochemical evaluation of renal mass revealed sarcomatoid RCC with rhabdoid feature.

KEY WORDS: Renal cell carcinoma · Sarcomatoid variant · Cardiac metastasis · Right ventricular outflow obstruction.

INTRODUCTION

Renal cell carcinoma (RCC) is a lethal cancer with aggressive behavior and a tendency to metastasis. Cardiac involvement along the renal vein and inferior vena cava (IVC) is a well-known situation during RCC progression, while in the absence of IVC involvement, clinically evident cardiac involvement is very rare, with only a few cases reported in the worldwide literature.¹⁾ Herein we report a case of a 61-year-old female of unusual type of RCC with right ventricular outflow tract (RVOT) mass. The RVOT mass was missed at preoperative echocardiography, but recognized at postoperative echocardiographic evaluation of cardiovascular collapse that occurred during surgical resection of RCC.

CASE

A 61-year-old female came to our hospital for left flank pain. Her abdominal computed tomography (CT) scan revealed about 9-cm sized exophytic heterogeneous enhancing mass at

the left kidney lower pole suggesting RCC (Fig. 1A). Multiple enlarged lymph nodes at the paraaortic, aortocaval, and retrocrural areas were also found, but there were no evidences of metastasis in the liver, spleen, pancreas, right kidney and great vessels. Bone scan showed no bony metastasis. Non contrast enhanced chest CT showed multiple metastatic lymph nodes at bilateral supraclavicular, mediastinal areas with small amount of pleural and pericardial effusion (Fig. 1B). Transthoracic echocardiographic (TTE) examination demonstrated normal heart size and function but scanty pericardial effusion without hemodynamic significance (Fig. 2, Supplementary movie 1). Although these findings indicated stage IV metastatic RCC, we decided to perform cytoreductive nephrectomy because the patient had good performance status with low metastatic burden at that time. During the operation, decreased blood pressure and subsequent cardiac arrest occurred. After cardiac resuscitation with extracorporeal membrane oxygenation circuit insertion, the patient was consulted to cardiology for the eval-

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uation of cardiac arrest. TTE showed markedly dilated right ventricle (RV) with D-shaped left ventricle suggesting RV pressure overloading and moderate amount of pericardial effusion

(Fig. 3A–D, Supplementary movie 2). And it also revealed a 5.5×3 cm sized echogenic mass nearly obstructing RVOT (Fig. 3E and F, Supplementary movie 3), which was attrib-

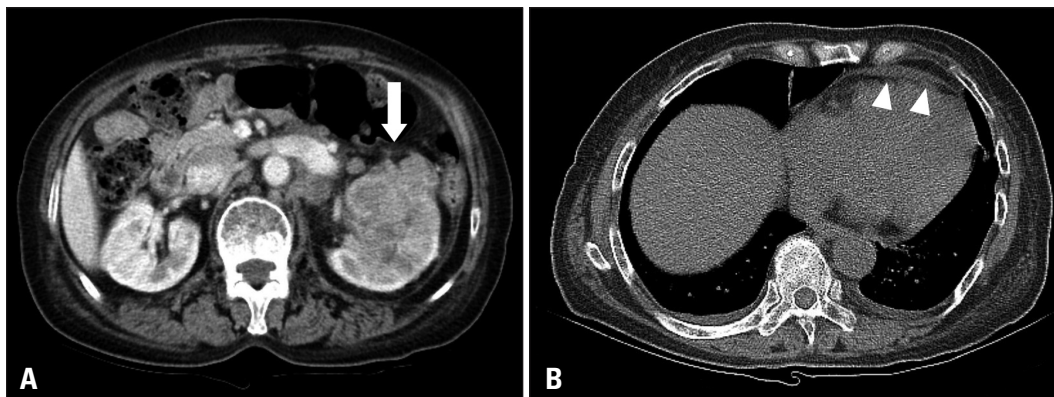


Fig. 1. Abdominal computed tomography (CT) revealed 9-cm sized left renal mass (arrow) (A). Chest CT showed scanty amount of pericardial effusion (arrowheads) (B).

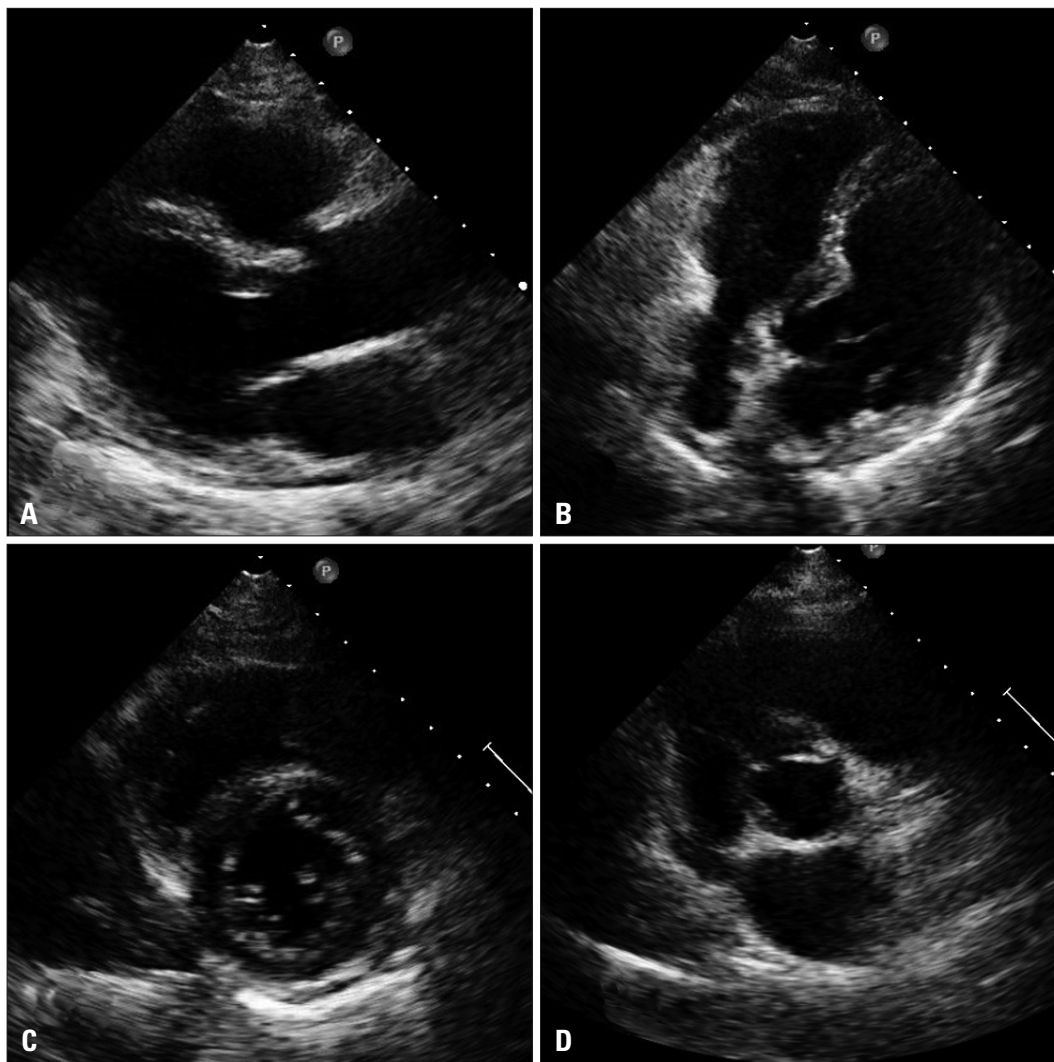


Fig. 2. Parasternal long axis view on transthoracic echocardiography showed normal left ventricle (LV) size and no evidence of right ventricular outflow tract mass (A). Modified four chamber view also showed normal LV and RV size with no evidence of intracardiac mass (B). In parasternal short axis view, no gross abnormality was observed (C and D). RV: right ventricle.

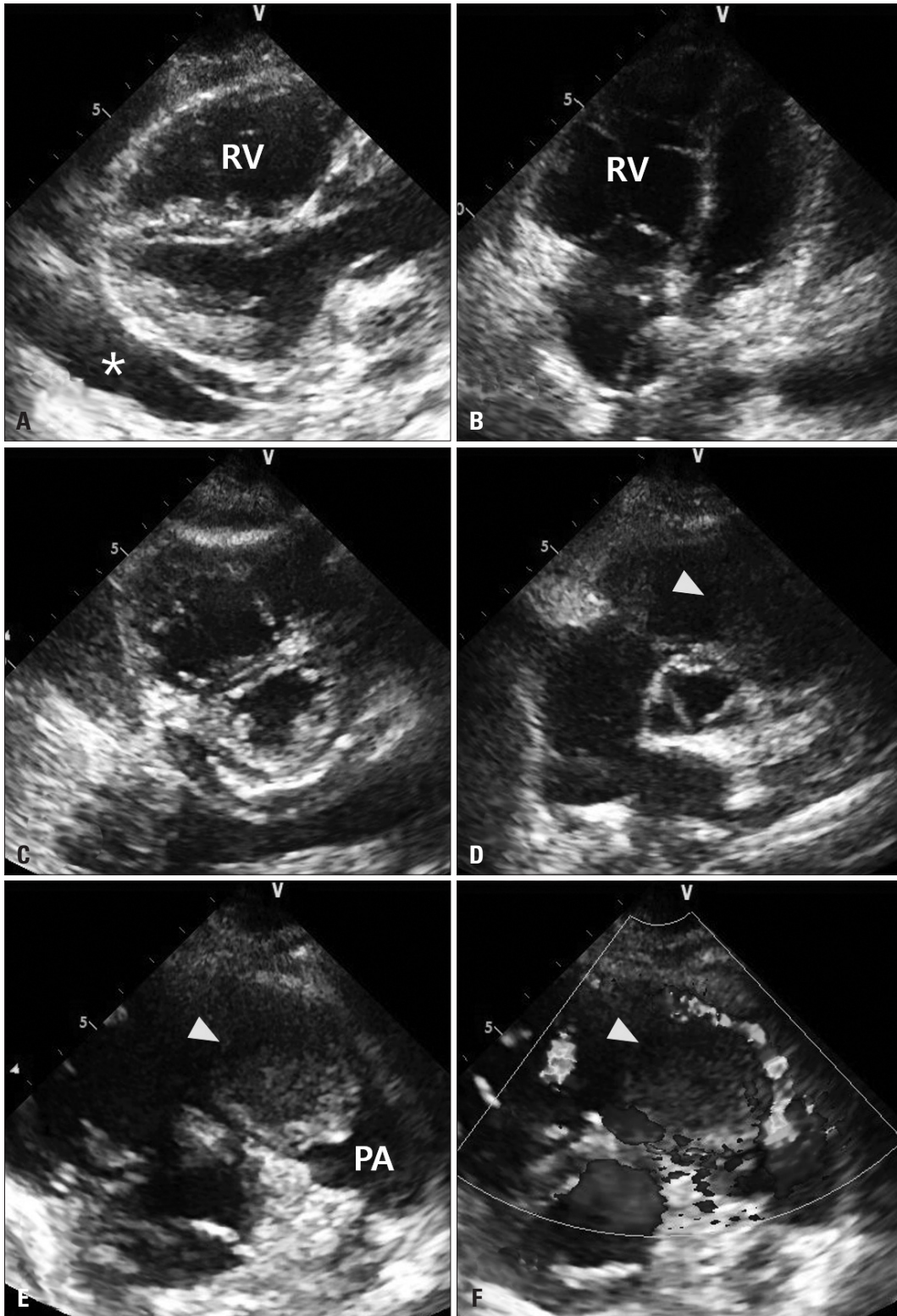


Fig. 3. Parasternal long axis view (A) and modified 4 chamber view (B) on transthoracic echocardiography revealed markedly dilated RV with moderate amount of pericardial effusion (asterisk). Parasternal short axis view showed D-shaped left ventricle (C). Parasternal short axis view of aortic valve level. In this view, right ventricular outflow tract (RVOT) mass (arrowhead) was hardly seen (D). Parasternal short axis view of RVOT level demonstrated 5.5 × 3 cm sized mass (arrowhead) nearly obstructing the RVOT (E). In this view, obstruction of blood flow by this RVOT mass (arrowhead) was well visualized under color Doppler image (F). RV: right ventricle, PA: pulmonary artery.

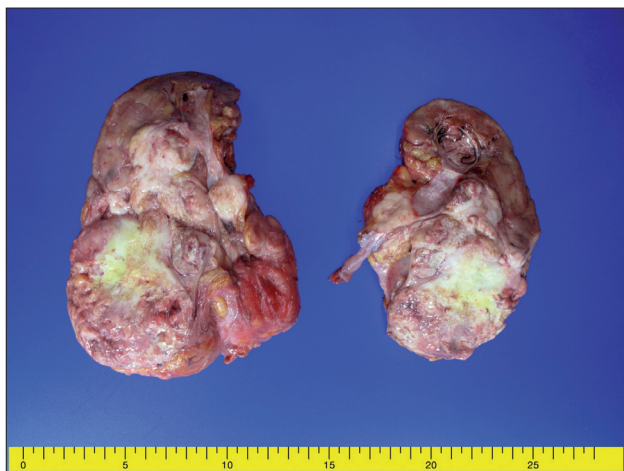


Fig. 4. The tumor showed an ill-defined, whitish, infiltrating mass with necrosis.

able to her hemodynamic instability. When we reviewed her previous TTE performed 1 month ago, we could recognize small RVOT mass which might be easy to be missed without attention (Fig. 2C and D). Resected left kidney revealed 11 × 10.5 × 8.4 cm sized whitish mass that showed infiltrating growth pattern and extensive necrosis (Fig. 4). Microscopic evaluation of the mass revealed malignant epithelioid cells. Most of them had rhabdoid morphology characterized by large vesicular eccentrically located nuclei, prominent nucleoli and abundant eosinophilic cytoplasm (Fig. 5A). Differential diagnosis included primary sarcoma, such as pleomorphic rhabdomyosarcoma and adult RCC with sarcomatoid and rhabdoid feature. Immunohistochemical stains showed diffuse strong positive for CD10 as well as vimentin and focal positive for pan cytokeratin, epithelial membrane antigen (EMA) and desmin (Fig. 5B–F). So we finally concluded that the renal mass

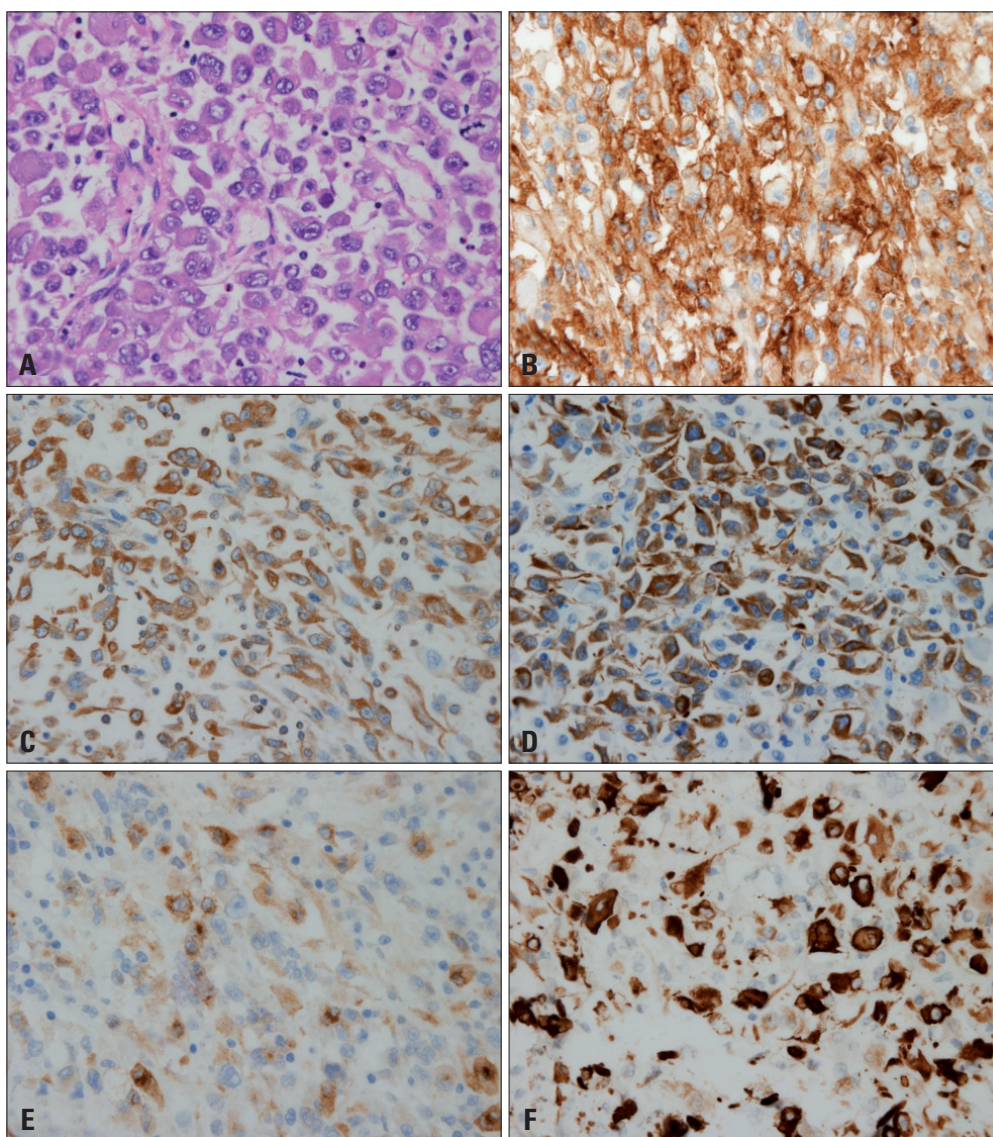


Fig. 5. The tumor composed of epithelioid tumor cells with rhabdoid feature. A: Hematoxylin and eosin staining (× 400). B: CD10 staining (× 400). C: Vimentin staining (× 400). D: Pan-cytokeratin staining (× 400). E: EMA staining (× 400). F: Desmin staining (× 400).

was sarcomatoid RCC with rhabdoid feature, a rare type of RCC. Although biopsy or operation of RVOT mass was not performed because her family refused another procedure or cardiac surgery, the mass was enough to be regarded as a secondary metastasis of RCC considering the very aggressive behavior of this type of RCC²⁾ and multiple metastases in lymph nodes of both thorax and abdominal cavity. The patient's condition deteriorated and she died 5 days after the operation.

DISCUSSION

Cardiac metastasis was shown to be present in 11% of patients who died of RCC.³⁾ The extension of a tumor into the renal vein or IVC as a luminal mass, with the growth along the caval wall into the right heart chambers, has been well documented as the most common mechanism of cardiac metastasis of RCC. Nevertheless, cardiac involvement of RCC without IVC involvement, as in our case, can present. Zustovitch et al.¹⁾ analyzed these cases and suggested another patterns of RCC metastasis to the heart; through the intrathoracic lymphatic systems, especially in the presence of disseminated disease. The latter is speculated as a metastatic pathway of our case, because in our case, intrathoracic lymph node metastasis and pericardial effusion was demonstrated.

A search of the literature revealed several cases of solitary RV metastasis of RCC without IVC involvement,¹⁾ but there has been only one case of metastasis confined to RVOT.⁴⁾ In this case, the metastasis was confirmed after 5 years after a radical nephrectomy and treated by echo-guided percutaneous coil embolization. To the best of our knowledge, our case is the first case that primary RCC and RVOT metastasis without involving IVC or right atrium were discovered almost simultaneously.

Another unique point of our case is the rare pathologic type of RCC, rhabdoid variants which represents dedifferentiation or divergent differentiation of RCC.⁵⁾ Recognition of this type has prognostic implications as it is associated with rapid progression and poor prognosis. Sarcomatoid RCC can metastasize unusual sites including breast,⁶⁾ colon and oropharynx.^{7,8)} But cardiac metastasis from sarcomatoid RCC is very exceptional.

RCC with rhabdoid feature is a rare type of RCC that car-

ries aggressive behavior with high metastatic potential and short survival time. When this type of pathology is confirmed, complete cardiac evaluation should be done in patients with RCC, especially in the presence of pericardial effusion. In addition, we should pay more attention to other cardiac and pericardial structures such as great vessels and mediastinum which can be unpredicted metastatic sites if signs of metastasis around these structures are present.

SUPPLEMENTARY MOVIE LEGENDS

Movie 1. Transthoracic echocardiogram parasternal short axis view at aortic valve level: no gross abnormality was seen.

Movie 2. Transthoracic echocardiogram parasternal short axis view at aortic valve level: right ventricular outflow tract mass was seen.

Movie 3. Transthoracic echocardiogram parasternal short axis view of the left ventricle at base level: huge mass obstructing the right ventricular outflow tract was seen.

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