

Unusual cause of right heart compression

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We report the case of a 77 year-old Caucasian woman with a past medical history of breast cancer who presented to our institution with a five-week history of progressive shortness of breath associated with abdominal distension, anorexia, change in bowel habit and cough. She had an ejection systolic murmur and a pansystolic murmur radiating to the

axilla, a raised jugular venous pressure, abdominal distension with shifting dullness and hepatomegaly 16 cm below the costal margin. Blood results revealed a normocytic anemia with deranged liver function tests, renal impairment and a raised C-reactive protein. Chest radiograph demonstrated a raised right hemidiaphragm (Fig. 1A). Transtho-

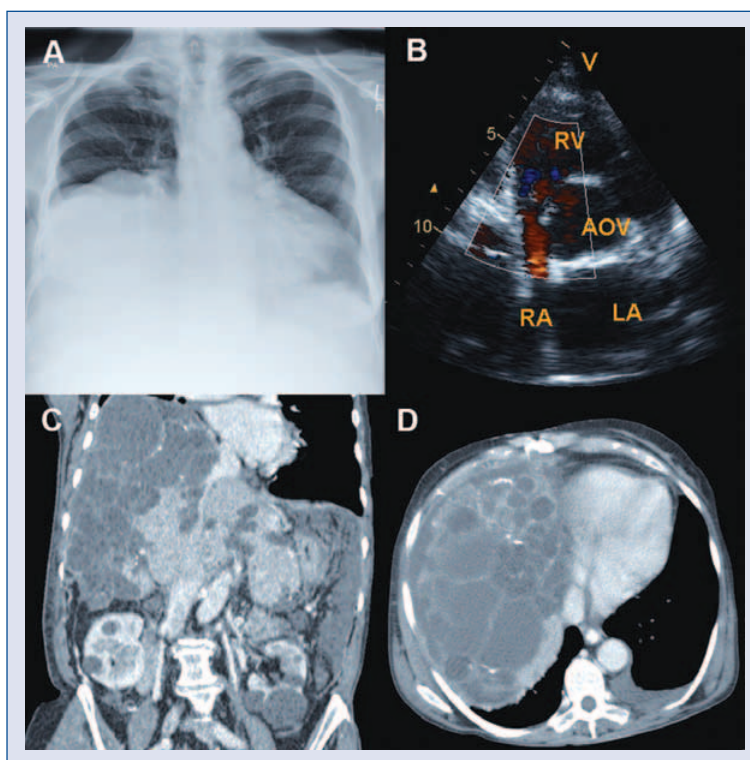


Figure 1. Chest radiograph demonstrating a raised right hemidiaphragm (A); transthoracic echocardiogram showing external compression of the right atrium and ventricle (B); coronal section of computed tomography demonstrating cysts of the liver, kidneys and pancreas (C); axial section of computed tomography demonstrating external hepatic compression of the right atrium and ventricle (D); RA — right atrium; LA — left atrium; AOV — aortic valve; RV — right ventricle.

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raphic echocardiography suggested external compression of the right atrium and ventricle, with high transtricuspid velocities (Fig. 1B). Computed tomography subsequently showed polycystic disease within the liver, pancreas and both kidneys, confirming a new clinical diagnosis of polycystic kidney disease (Fig. 1C). The cystic enlargement of the liver had caused a degree of compression of the right heart chambers and inferior vena cava (Fig. 1D) as well as a raised right hemidiaphragm. In addition, a cystic lesion arising from the left adnexa was noted. Subsequent ascitic fluid cytology was suggestive of ovarian metastatic adenocarcinoma in keeping with a markedly raised Ca125.

Unfortunately, the patient sustained a sudden cardiac arrest and passed away in the intensive care unit after initial resuscitation. Massive pulmonary embolism or decompensation secondary to further right heart compression was suspected, but the family declined a post-mortem examination. A first-degree relative was discovered to have cystic enlargement of the liver and a renal cyst, and was referred to the Department of Genetics for further investigation.

The liver is the commonest site of extrarenal manifestations in polycystic kidney disease [1].

There has been only one case reported in the literature of refractory hypotension and edema caused by right atrial compression in a woman with polycystic kidney disease [2]. However, the novelty in our case is the extent of the cystic hepatomegaly causing compression of both right heart chambers, and the presentation of dyspnea rather than hypotension in a patient not previously known to have polycystic kidney disease. This case illustrates the growth potential of cystic hepatic enlargement in polycystic kidney disease.

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References

1. Pirson Y. Extrarenal manifestations of autosomal dominant polycystic kidney disease. *Adv Chronic Kidney Dis*, 2010; 17: 173–180.
2. Lasic LB, DeVita MV, Spiegel PJ, Marino ND, Mellow E, Michelis MF. Refractory hypotension and edema caused by right atrial compression in a woman with polycystic kidney disease. *Am J Kidney Dis*, 2004; 43: e13–e17.