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Extracardiac Compression of Right Atrium by Hepatic Cysts in a Patient with Polycystic Kidney Disease

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Background: We describe a case of right heart compression by severe cystic liver disease, which is rarely reported in patient with polycystic kidney disease.

Methods: A 50 year-old male with background history of autosomal dominant polycystic kidney disease (ADPKD), hypertension and end-stage renal disease on hemodialysis presented to us with sepsis due to infected renal cysts. A computed tomography (CT) scan of the abdomen revealed polycystic kidneys and massively enlarged liver containing innumerable cysts involving all the segments resulting in elevated right hemidiaphragm and compression of right heart (Figure). Transthoracic echocardiography (TTE) suggested extracardiac compression of right atrium (RA) by hepatic cysts resulting in impaired filling (Figure). Despite the above findings, the patient was asymptomatic and did not show any sign of hemodynamic compromise including during his regular hemodialysis. He was subsequently followed-up under multidisciplinary care formed by nephrologist, cardiologist and hepatobiliary surgeon. He remained stable and a repeat TTE performed a year later showed no significant disease progression.

Results: Hepatic cysts are a common extrarenal manifestation of ADPKD and risk factors such as increasing age, female gender, multiple pregnancies and exogenous estrogen can result in progression into severe cystic liver disease. Most patients are asymptomatic but severe cyst burden has been reported to cause hemodynamic complications including inferior vena cava compression, hepatic venous outflow obstruction and portal hypertension. Direct compression of right heart by hepatic cysts has been rarely reported, and was described to be associated with refractory hypotension in the presence of significantly impaired right ventricular filling. Therapeutic options for symptomatic patients include cyst aspiration and sclerotherapy, cyst fenestration, transcatheter arterial embolization or surgical interventions including cyst resection and liver transplantation.

Conclusion: RA compression by hepatic cysts is a rare feature in ADPKD and can potentially lead to cystic and hemodynamic complications which will require follow up by a multidisciplinary team.

Keywords: atrial compression, liver cysts, polycystic kidney disease