Letters to Editor

Post-operative refractory hypotension due to right heart compression by hepatic cysts in a patient with autosomal dominant polycystic kidney disease

Sir,

A 46-year-old male, diagnosed case of autosomal dominant polycystic kidney disease (ADPKD) type 1 since last 23 years was transferred to our intensive care unit (ICU) for management of respiratory distress and hypotension. The patient had undergone left nephrectomy 2 weeks ago. Prior to this surgery, his hypertension was adequately controlled with regular enalapril therapy and he was on biweekly maintenance hemodialysis for his advanced renal failure. Furthermore, at this time, echocardiography (ECHO) had revealed left ventricular (LV) hypertrophy with LV ejection fraction (EF) of 60% and right atrial compression by a hepatic cyst.

On being transferred to ICU, patient was noted to be conscious but confused, tachycardic with heart rate of 120/min and had a blood pressure of 80/60 mmHg and central venous pressure of 10 mmHg. He had a serosanguinous fluid oozing from the nephrectomy wound along with grossly distended abdomen and bilateral pedal edema. Investigations revealed leukocytosis (18,000/cmm) with deranged renal function, but with normal liver functions. Keeping in view with the diagnosis of septic shock, he was managed with mechanical ventilation, broad spectrum antibiotics, renal replacement therapy, 20% albumin and vasopressor infusion (noradrenaline to a maximum of $0.5 \,\mu g/$ kg/min). Despite an initial recovery from sepsis the requirement for noradrenaline remained high along with elevated (18 mmHg) intra-abdominal pressures as measured by intra-urinary-bladder closed-system repeated-measurement technique.^[1] Subsequent non-contrast computed tomography scan of abdomen showed grossly enlarged polycystic liver [Figure 1] with moderate ascites. A therapeutic ascitic fluid tapping of 1.8 L was carried out to reduce the abdominal pressure. The ECHO showed a dilated LV with LV EF of 30% and right atrial and ventricular compression by hepatic cyst [Figure 2]. Hepatic resection along with right partial nephrectomy was planned, but the family members did not give consent for the procedure. Eventually, patient developed multi-drug-resistant abdominal infection and succumbed to his illness.

Two previous reports of similar hemodynamic compromise due to compressive effect of the polycystic liver in ADPKD could be located.^[2,3] The number and size of cysts in the liver increase with age, multiple pregnancies and due to effect of estrogen.^[3,4] Both patients reported previously were females. One patient was managed by right nephrectomy as patient was high-risk for hepatectomy. And another patient recovered spontaneously after abdominal blunt trauma (cyst size decreased from 23 cm to 11 cm within a few days).^[2,3] Our patient was the first reported male patient with such post-operative complications.

Therapeutic options available in patients with symptomatic liver cysts includes percutaneous cyst aspiration along with



Figure 1: Non-contrast computed tomography of abdomen showing massively enlarged liver (arrowed) with multiple cysts

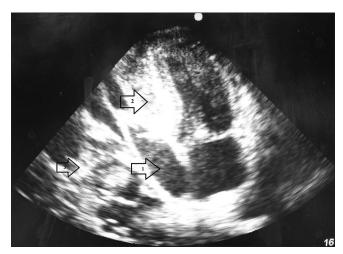


Figure 2: 2D echocardiogram apical 4 chamber view showing right atrium (1) and right ventricle (2) compressed by the hepatic cysts (3)

sclerotherapy, cyst fenestration; percutaneous portal venous stenting liver resection and liver transplantation.^[4] Cyst aspiration and fenestration of the cysts was considered in our patient, but deferred due to inaccessibility of the critically affected area of the liver and high chances of cyst recurrence post-aspiration.^[2] We believe that any compression on right heart as described in pre-operative ECHO should be taken seriously. Hugely dilated and cystic left kidney might force surgeons to do a left nephrectomy to provide symptomatic relief. Retrospectively, it appears that in our case, refractory hypotension could have been possibly avoided if bilateral nephrectomy or partial hepatectomy^[2] had been carried out intra-operatively instead of left nephrectomy only. It could create space for the liver to descend in abdomen and produce less cardiac compression.

Tanmoy Ghatak, Afzal Azim, Arun Sharma, Samir Mahindra¹

Departments of Critical Care Medicine, 'Gastro Medicine, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

> Address for correspondence: Dr. Tanmoy Ghatak, C/o Dr. Tarun Kumar Ghatak, Rammohan Pally, Arambagh, Hooghly - 712 601, West Bengal, India. E-mail: tanmoyghatak@gmail.com

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