

# Why Did The Patient with A History of ADPKD Faint? The Giant Liver Cyst Explained Everything: A Case Report

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## Abstract

Syncope is a common complaint in emergency clinics, but the symptoms of syncope are quite comprehensive. In this study, we are going to present a case of a non-parasitic giant liver cyst that caused compression of the inferior vena cava with the right atrium in a 47-year-old female patient with a history of autosomal dominant polycystic kidney disease who applied to the emergency department due to syncope. In the examinations performed in the emergency department, we detected a giant liver cyst pressing on the inferior vena cava, right atrium, and ventricle of the heart, which prevents venous return. Sclerotherapy with catheterization was applied to the non-parasitic giant liver cyst and the drainage catheter was kept in the cyst cavity for one week to prevent an early recurrence.

**Key words:** Giant hepatic cyst, syncope, orthostatic hypotension, heart compression, inferior vena cava compression

## Introduction

Syncope is a sudden and brief loss of consciousness associated with a loss of postural tone following spontaneous recovery. The general pathophysiology of syncope types consists of a sudden decrease or short-term interruption of cerebral blood flow<sup>1</sup>.

Causes of syncope range from non-serious to potentially fatal. The most common causes of syncope are cardiovascular pathologies, orthostatic hypotension and reflex also known as neural-mediated hypotension<sup>1</sup>.

Initial evaluation, a detailed history, detailed physical examination, electrocardiogram (ECG), blood tests are helpful in identifying the underlying cause of syncope<sup>1</sup>.

In this case, we aimed to present an autosomal dominant polycystic kidney disease (ADPKD) patient who developed syncope due to the giant cyst of the liver compressing the heart and inferior vena cava (IVC).

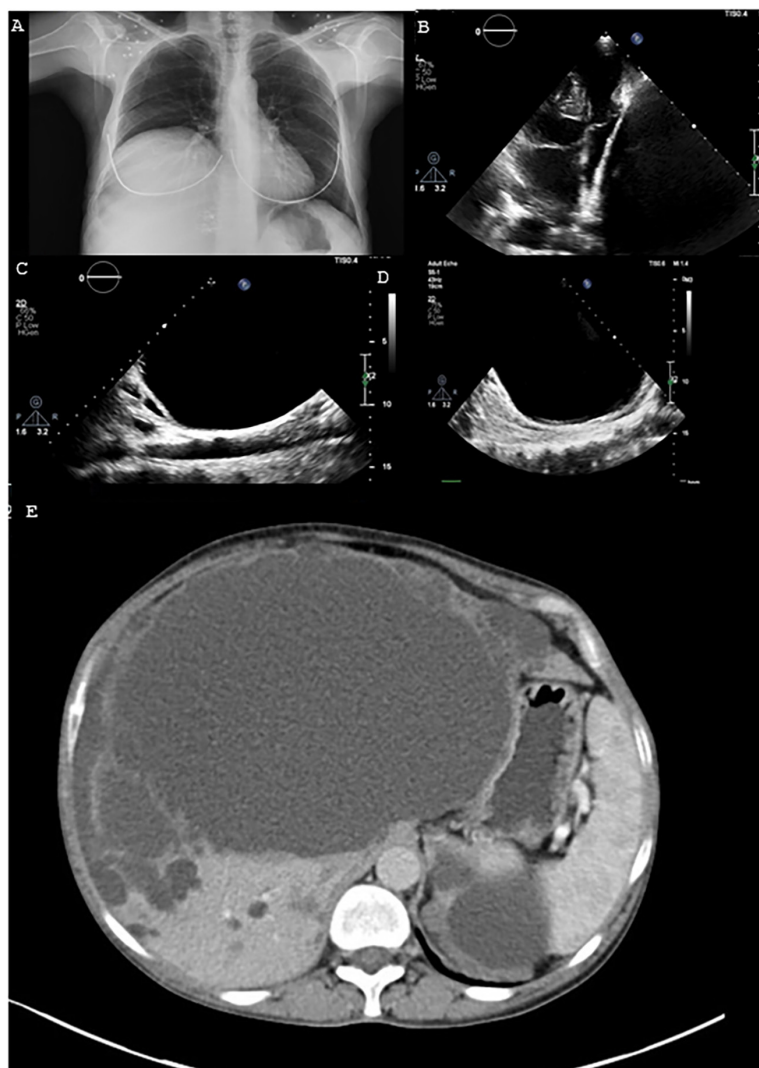
## Case

A 47-year-old young female patient was brought to the emergency department due to syncope when she suddenly stood up. On arrival, the patient's vital signs were: blood pressure of 95/55 mmHg, oxygen saturation of 97% on room air, respiratory rate of 20 breaths/minute, heart rate of

122 beats/minutes, temperature of 36,7 degrees Celsius, fingertip blood sugar is 96 mg/dL. There was no symptoms of fever, chest pain, shortness of breath, nausea, and vomiting. At the time of syncope, a friend of the patient who was next to her prevented her from falling to the ground.

It was reported that the patient had a history of ADPKD, undergoes routine dialysis 3 times a week and has complaints of bloating, decreased food intake due to early satiety and loss of appetite for the last month. It was stated that her last dialysis treatment was 1 day prior to fainting. ECG shows sinus tachycardia with 122 beats/minute without ST, T wave change. Her neurological examination was normal. On abdominal examination, a palpable mass was palpated in the abdomen.

In the examinations, there were no scleral icterus or significant swelling in the lower extremities. Her digital rectal examination was normal. Necessary laboratory examinations were requested. Laboratory findings were Glucose 96 mg/dL (74-106), WBC 5.16 10<sup>3</sup> / mm<sup>3</sup> (4 - 10.5), NEU 3.92 10<sup>3</sup> / mm<sup>3</sup> (1.82 - 7.42), LYM 1,4 10<sup>3</sup> / mm<sup>3</sup> (0.85-3), Blood Urea Nitrogen 111 mg / dL (8 - 20), Creatinine 6.44 mg / dL (0.81 - 1.44), Alanine aminotransferase (ALT) 6 U / L, Aspartate transaminase AST- (SGOT) 4 U / L, CRP 14 mg / L (0 - 5), potassium (K): 4.71 mmol / L (3.5-5.0), sodium (Na): 136 mmol / L (138 - 145), Troponin T: 24 ng \ L (0-14) calcium: 8.87 mg / dL (8.5-10.5), pH 7.36(7.35-7.45), PaCO<sub>2</sub> 40 mm Hg(35-45), PaO<sub>2</sub> 90mm Hg (80-100), HCO<sub>3</sub> 20 mmol/L(22-26), Lactate 1.4 mmol/L(0.5-2)



**Figure 1:** (A) X-ray image of lungs. (B) TTE imaging of compression of the RA. (C) Compression of the giant cyst in the liver to the inferior vena cava during inspiration. (D) Compression of the giant cyst in the liver to the inferior vena cava during expiration. (E) CT axial plane at the largest cyst diameter level: 21 x 15.5 cm.

The patient was administered a physiological saline solution. Cranial tomography and diffusion MRI was found to be normal. Elevation in the right diaphragm was detected on chest X-ray. (Figure 1: A) Bedside echocardiography was performed to investigate the cause of syncope.

In our patient with normal ejection fraction, an extracardiac mass compressing the right atrium and right ventricle was observed (Figure 1: B) Mass turned out to be a giant liver cyst and severe IVC compression ( $\geq 70\%$ ) was detected on abdominal USG. (Figure 1: C-D). Venous Doppler was found to be normal in the bilateral lower extremities. Intravenous opaque thorax and abdomen CT showed along with many small cysts in the liver, a giant liver cyst of 21x15.5 cm putting pressure on inferior vena cava, stomach, diaphragm and mild pressure on the gallbladder. The common bile duct and intrahepatic bile ducts appears normal. (Figure 1: E) There were multiple cysts in both kidneys. Pulmonary embolism and thrombus in the inferior vena cava were not observed.

No significant changes were detected in cardiac marker and ECG follow-up. The compression of the giant liver cyst

was suspected as the underlying cause for syncope. The patient was hospitalized for the application of drainage of the symptomatic giant liver cyst.

## Discussion

Syncope is a common main complaint at emergency clinic admission. Causes of syncope range from non-serious to potentially fatal ones<sup>2</sup>. Cardiovascular pathologies, orthostatic hypotension and reflex also known as neural mediated are the most known causes of syncope<sup>1</sup>.

In our case, a giant cyst in the liver, which was detected by ultrasonography in the emergency department, was compressing the inferior vena cava and right atrium ventricular. When the patient abruptly stood up, a giant liver cyst that compressing the inferior vena cava reduced venous return hence syncope occurred due to orthostatic hypotension. No pathological findings explaining the clinic were detected in the laboratory examination. Gastrointestinal bleeding was

not considered in the foreground with the presence of normal stool contamination in digital rectal examination. It was thought that a possible cardiac dysrhythmia that could cause syncope would be unlikely due to obviously appearing giant hepatic cyst that prevented venous return. In a case report similar to our case, syncope developed due to the compression of the giant cyst in the liver on the heart and inferior vena cava<sup>3</sup>.

A case of Polycystic Kidney Disease (PKD) with right-sided heart failure due to liver cysts pressing on the right atrium have been described in the related literature<sup>4</sup>. Our patient had both right atrial and right ventricular compression, but there was no evidence of right heart failure.

Most simple cysts in the liver are usually detected incidentally on imaging because of their asymptomatic behavior. The most commonly encountered symptom in this patient group is abdominal pain<sup>5</sup>. Interestingly, our patient did not have severely disturbing abdominal pain. She stated that she felt the bulk in her abdomen, but she did not seek treatment at the hospital before because she did not have any disturbing pain.

Large cysts in the liver show obvious symptoms such as abdominal bloating, fullness, early satiety, nausea, and vomiting. The most common complications of liver cysts are rupture, infection, obstructive jaundice, cyst bleeding, portal vein occlusion with splenic varices, and inferior vena cava thrombosis<sup>6</sup>.

The reason for the patient's early satiety and decreased oral intake is due to the compression of the stomach by the giant cyst detected in the liver. No thrombus was detected in the inferior vena cava and pulmonary artery in Contrast CT. There was no evidence of obstructive jaundice infection. One of the case-control study of the patients with APDKD found that the prevalence of IVC compression by kidney or liver cysts was dramatically higher in women older than 40 years and in women<sup>6</sup>.

This is due to the fact that vena cava compression occurs in women who are more likely to have more and larger liver cysts than men, possibly due to an estrogen-related mechanism<sup>7</sup>. The fact that our patient was over 40 years old, and female supports this hypothesis.

In addition, in this study, it was found that some of the cases with IVC compression worsen progressively. In addition, no regression was detected in IVC compression in any of them. Therefore, symptomatic patients require intervention. Percutaneous aspiration or surgical intervention is required<sup>8</sup>.

For symptomatic non-parasitic liver cysts, the recommended treatment modalities are open deroofting and percutaneous sclerotherapy, as simple drainage often results in recurrence<sup>9</sup>. In our case, sclerotherapy was performed with catheterization and the drainage catheter was kept in the cyst cavity for a week to prevent an early recurrence.

In conclusion, IVC compression by hepatic cysts formation in APDKD patients is an important vascular complication. This should be kept in mind when evaluating APDKD patients in the emergency department.

## Conclusion

Due to the irregular follow-ups, the size of the cysts formed in the kidney and other organs of our patient with APDKD was not followed up, that's why a liver cyst of that size wasn't diagnosed earlier. In patients diagnosed with APDKD, ultrasound should be performed at regular intervals to monitor the size of the possible cysts. When evaluating APDKD patients in the emergency department, we suggest that patients should be evaluated quickly with bedside ultrasound for pathologies caused by cysts developing in the kidneys and other organs.

## References

1. Brignole M, Moya A, de Lange FJ, et al. 2018 ESC Guidelines for the diagnosis and management of syncope. *Eur Heart J*. 2018;39(21):1883-1948. doi:10.1093/eurheartj/ehy037
2. Olde Nordkamp LR, van Dijk N, Ganzeboom KS, et al. Syncope prevalence in the ED compared to general practice and population: a strong selection process. *Am J Emerg Med*. 2009;27(3):271-279. doi:10.1016/j.ajem.2008.02.022
3. Belyavskiy E, Tadic M, Hassfeld S, Pieske B. A Giant Hepatic Cyst: A Rare Cause of Syncope. *Can J Cardiol*. 2018;34(9):1234.e1-1234.e2. doi:10.1016/j.cjca.2018.05.014
4. Algin A, Özdemir S, Sariaydin M, Erdoğan MÖ, Inan I. Cardiac compression of a hepatic cyst in polycystic liver disease: A rare cause of hemodynamic instability. *Turk J Emerg Med*. 2020;20(2):93-96. Published 2020 Feb 26. doi:10.4103/2452-2473.279558
5. Plard L, Guedin P, Le Pennec V, Chiche L. Kystes hépatiques dits "kystes biliaires du foie." [Hepatic cysts: diagnosis and management]. *J Chir (Paris)*. 2008;145(3):217-225. doi:10.1016/s0021-7697(08)73749-9
6. Yin X, Blumenfeld JD, Riyahi S, et al. Prevalence of Inferior Vena Cava Compression in ADPKD. *Kidney Int Rep*. 2020;6(1):168-178. Published 2020 Nov 1. doi:10.1016/j.ekir.2020.10.027
7. van Aerts RMM, Bernts LHP, Gevers TJG, et al. Estrogen-Containing Oral Contraceptives Are Associated With Polycystic Liver Disease Severity in Premenopausal Patients. *Clin Pharmacol Ther*. 2019;106(6):1338-1345. doi:10.1002/cpt.1553
8. Hansen P, Bhojru S, Legha P, Wetter A, Way LW. Laparoscopic treatment of liver cysts. *J Gastrointest Surg*. 1997;1(1):53-60. doi:10.1007/s11605-006-0010-1
9. Vardakostas D, Damaskos C, Garmpis N, et al. Minimally invasive management of hepatic cysts: indications and complications. *Eur Rev Med Pharmacol Sci*. 2018;22(5):1387-1396. doi:10.26355/eurrev\_201803\_14484.