Forensic Science International 262 (2016) e5–e8

Contents lists available at ScienceDirect

Forensic Science International

journal homepage: www.elsevier.com/locate/forsciint

Case report

Fatal liver cyst rupture in polycystic liver disease complicated with autosomal dominant polycystic kidney disease: A case report

Fang Tong [a], Yue Liang [a], Lin Zhang [a], Wenhe Li [a], Peng Chen [b], Yijie Duan [a], Yiwu Zhou [a,*]

[a] Department of Forensic Medicine, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, PR China
[b] Department of Forensic Biology, West China School of Preclinical and Forensic Medicine, Sichuan University, Chengdu 610041, PR China

ARTICLE INFO

Article history:
Received 13 November 2015
Received in revised form 20 March 2016
Accepted 21 March 2016
Available online 30 March 2016

Keywords:
Forensic pathology
Autosomal dominant polycystic kidney disease
Polycystic liver disease
Genetic analysis

ABSTRACT

A 59-year-old man was struck in the abdomen and later presented to the emergency room. His blood pressure dropped and eventually died 16 h post trauma and just before emergency exploratory laparotomy. Autopsy revealed two polycystic kidneys and a giant polycystic liver with two ruptures. Blood (2225 g) was observed in the peritoneum and the body-surface injury was minor. Genetic testing was performed to confirm that the man had an autosomal dominant polycystic kidney disease (ADPKD) complicated by polycystic liver disease (PLD). Autopsy, histopathology and medical history showed that the cause of death was the ruptures of liver cysts due to trauma. In this communication, we describe a fatal case and hope to increase awareness and recognition of PLD and ADPKD. We also wish to indicate that due to the fragile condition of liver cysts, trauma should be considered even if the body-surface injury is minor in fatal cases of PLD patient with a traumatic history.

1. Introduction

Polycystic liver disease (PLD) is a rare anomaly caused by malformation of biliary duct in the liver [1–3]. The disease was first reported in 1856 in combination with autosomal dominant polycystic kidney disease (ADPKD) [4]. There are three kinds of PLD entities: ADPKD, autosomal dominant polycystic liver disease (PCLD) and Von Meyenburg complexes [5,6], all of which have different pathologies and pathogenesis. Investigators have shown that PLD is clinically silent in early stage and approximately 50% of PLD patients with massive hepatomegaly will suffer from severe complications such as cyst hemorrhage, cyst rupture, and/or a cyst infection in the final stages [4,7,8].

Autopsy and histopathology findings in this particular case showed that the PLD of the deceased was associated finding of ADPKD. ADPKD is a life-threatening disease characterized by fluid-filled renal cysts, which could lead to renal failure due to the progressive enlargement of cysts [9]. Complications such as PLD, intracranial aneurysms, aortic root dilatation and aneurysms, mitral valve prolapse, hypertension and abdominal wall hernias are well described in literature [3]. Genetically, a certain mutation in PKD1 and/or PKD2 is associated with ADPKD [3,9].

Here, we present a case of a PLD patient who died following an impact in the abdomen that disrupted the liver cysts. A postmortem genetic analysis was performed to further confirm the diagnosis of ADPKD. To a certain extent, the minor body-surface injury of the deceased complicated the evaluation of the disease, trauma and death.

2. Case report

A 59 year-old man was kicked in the abdomen in a fight at 12:00 noon. He fell to the ground and was sent to the emergency room 2 h later. His blood pressure dropped to 75/65 mm Hg at 14:20. Blood work revealed WBC at 11.8×10^9/L, RBC at 3.38×10^{12}/L and HB 98 g/L. Ultrasonography showed significant fluid accumulation in the peritoneum. He received a conservative treatment initially and was administered medications to maintain a normal blood pressure. At 2:00 a.m. of the next day (14 h post-trauma) his blood pressure suddenly dropped to 85/52 mm Hg and abdominal distention was quite obvious. Abdominal tenderness and positive ascites signs were evident. It was decided that the patient required an emergency exploratory laparotomy to stop abdominal bleeding. At 2:52 a.m. (15 h post-trauma), his heart stopped just before the surgery. Ultimately, he was announced dead at 4:17 a.m. despite

* Corresponding author. Tel.: +86 27 83657925/cell phone: +86 13707160935; fax: +86 27 83692638.
E-mail addresses: fytongfang@gmail.com (F. Tong), yueyueashin@gmail.com (Y. Liang), lyre2008@126.com (L. Zhang), 18707165089@163.com (W. Li), 276538406@qq.com (P. Chen), gerichtsarzt@hotmail.com (Y. Duan), zhouyiwu@outlook.com (Y. Zhou).

http://dx.doi.org/10.1016/j.forsciint.2016.03.045
0379-0738/

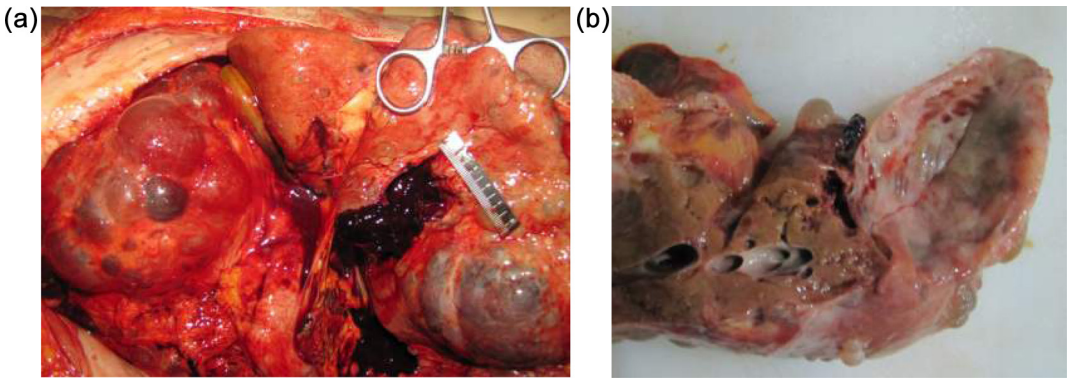

Fig. 1. a. The rupture on left lobe of the liver with clotted blood around it. b. The rupture on left lobe of the liver with clotted blood around it.

all treatments. Past medical history revealed that the deceased had a history of chronic renal failure and hypertension and once had an operation to repair an abdominal wall hernia.

A forensic autopsy was performed 2 days after death. The body was 157.0 cm long. The abdomen was obviously enlarged and there was an abdominal wall hernia, 6.5 cm in diameter with a surgical scar right beside it. External examination did not show any lethal injury.

The liver was large, 3347 g in weight and 32.0 cm × 21.0 cm × 12.0 cm in volume and contained many cysts. The liver edge was extended below the costal margin and there was a large quantity of blood, 2255 g, mainly beneath the liver in the peritoneum. The surface of the liver was irregular, with many cysts throughout. The cysts varied in size, were thin-walled and filled with either brown or yellow fluid. We found a giant ruptured cyst, 11.0 cm × 7.0 cm × 5.0 cm in size, on the left lobe of the liver with an 11.0 cm long rupture on the visceral surface and a large volume of clotted blood adhering around the rupture (Fig. 1a and b). There were smaller ruptured cysts, 5.0 cm × 3.0 cm × 2.5 cm, near the boundary of the two lobes. The length of the second rupture was 2.5 cm, and with parenchymal hemorrhage. Microscopically, the cysts were lined with columnar, cuboidal or flattened epithelium and surrounded by a scant of fibrous tissue with infiltration of inflammatory cells. Hemorrhage was observed in the tissue around the rupture (Fig. 2). Most of the parenchyma was normal.

The two kidneys were both irregularly shaped with many cysts (Fig. 3). The left one was 650 g in weight and 15.0 cm × 9.0 cm × 8.5 cm in size, and the right, 2000 g in weight and 22.0 cm × 12.0 cm × 11.0 cm in size. Histopathological examination showed numerous cysts surrounded by cuboidal epithelium and hyperplasia of fibrous connective tissue in the renal interstitial tissue (Fig. 4).

The heart was 610 g in weight and coronary atherosclerosis was observed in the LAD, LC and RM. The most severe atheromatous plaque was in the LAD, occupying more than 75% of the lumen. Microscopic examination showed the left ventricle and interventricular septum had fibrosis.

Most of the main organs of the deceased were anemic in appearance. No other significant pathological changes were found.

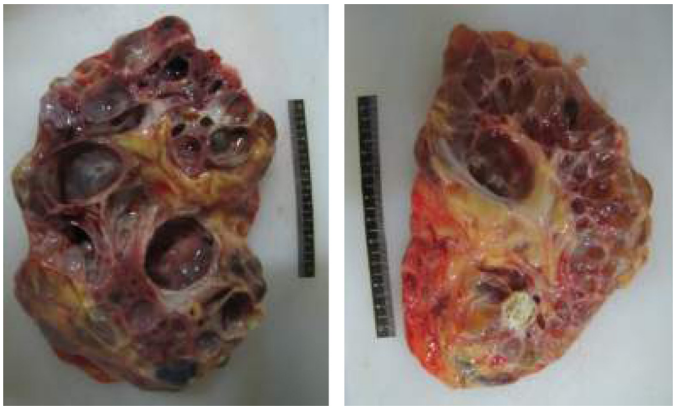

Fig. 3. The two kidneys were both irregularly shaped with many cysts.

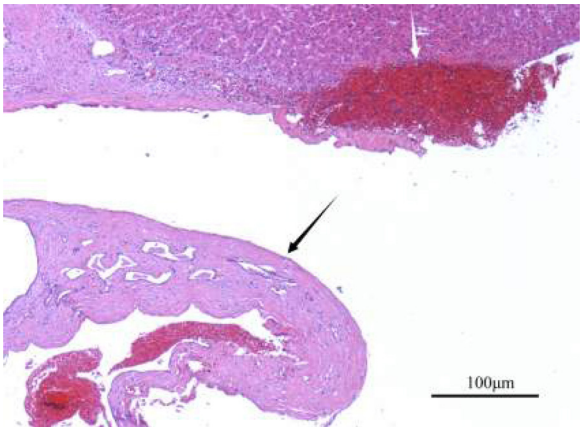

Fig. 2. The clotted blood and rupture of the liver (white arrow), liver cyst (black arrow)

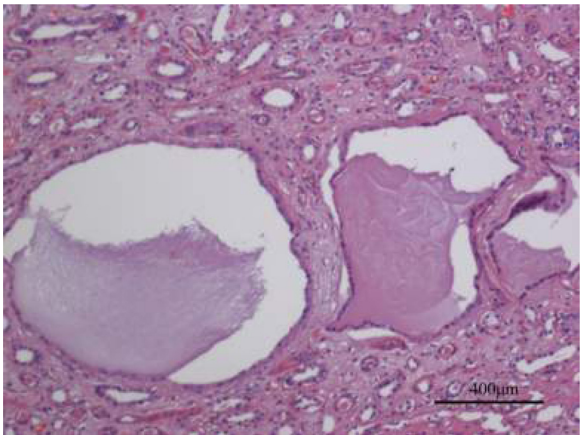

Fig. 4. Kidney cysts on microscope HE × 100