Case Report

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Polycystic Liver Disease

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ABSTRACT

Keywords: Polycystic Liver Disease; Autosomal Dominant Polycystic Kidney Disease; Gigot Classification; Fenestration; Hepatectomy

Clinical Image

We report the case of a 46-year-old woman, known to have polycystic hepatorenal disease, who consulted for increased abdominal distension and secondary respiratory distress. On examination, her abdomen was grossly distended, with multiple cystic nodules in the supraumbilical region (Figure 1).

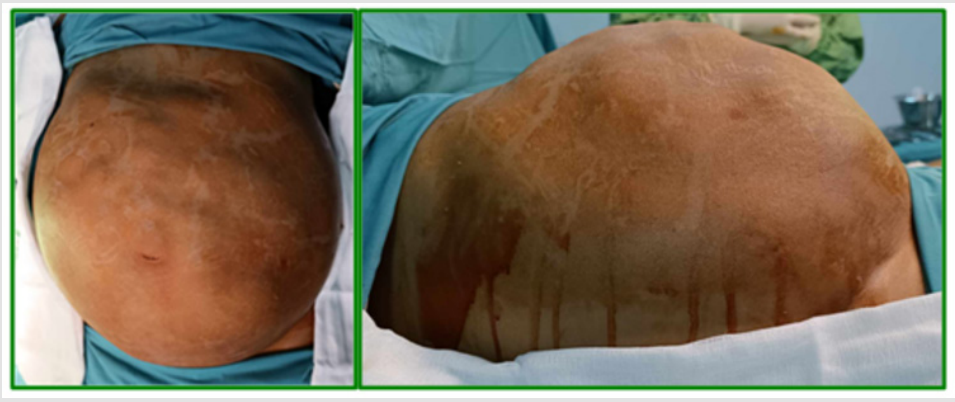

Figure 1: Distended abdomen with multi-nodular appearance.

Laboratory Tests Showed Normal Renal and Hepatic Function

Abdominal CT showed an enlarged liver, riddled with simple cystic lesions, classified as type III according to the GIGOT classification. Both kidneys showed numerous small cortical serous cysts. The patient was a candidate for liver transplantation. Given the long waiting time and shortage of transplants, surgical fenestration was decided to be performed. Intraoperatively, the liver had a multi-cystic appearance, with liver parenchyma totally involuted on the left (Figure 2). She underwent fenestration of the cysts and a left lobectomy (Figure 3), which resulted in an accidental wound of the middle hepatic vein, controlled by suture. The patient was transfused and managed intraoperatively using vasoactive drugs. She died of diffuse intravascular coagulation immediately postoperatively.

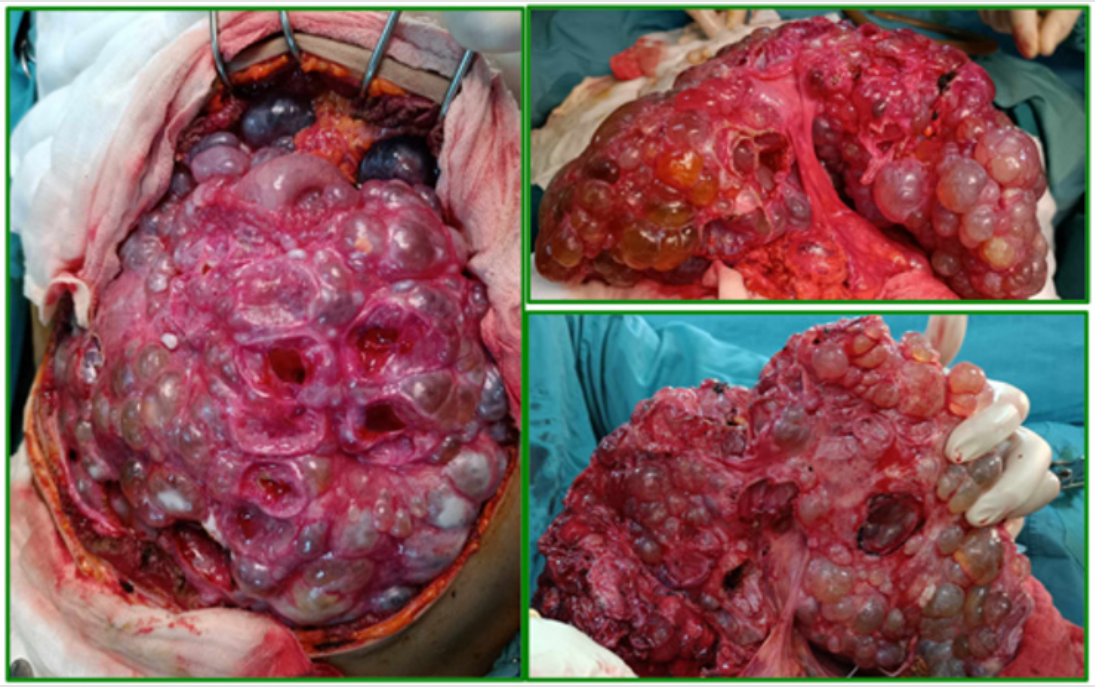

Figure 2: Liver riddled with simple cysts intraoperatively.

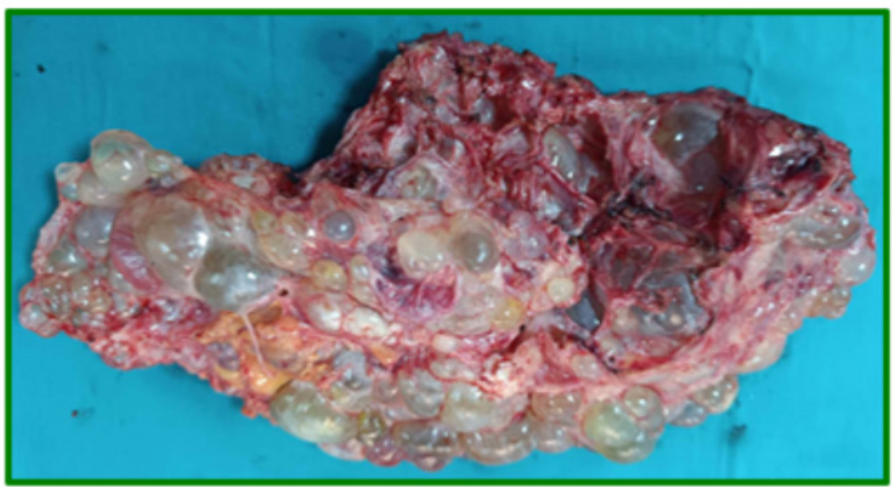

Figure 3: Surgical specimen of resected left hepatic lobe.

Polycystic liver disease is a rare hereditary disorder defined by the presence of multiple diffuse cysts in the liver. It may be isolated, or accompany autosomal dominant or autosomal recessive polycystic kidney disease [1]. Autosomal dominant polycystic kidney disease (ADPKD) is the most common monogenic genetic kidney disease. Polycystic liver disease is the most frequent extrarenal manifestation of ADPKD, affecting 94% of patients [2]. The number and size of cysts and the proportion of functional liver parenchyma remaining are the determinants of GIGOT classification. It has divided polycystic liver disease into three types, of which type III has the poorest prognosis [3]. The treatment of hepatic cysts may include percutaneous aspiration, fenestration, lobectomy or hepatectomy, or even liver transplantation [3].

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