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Case Report **Hepato-Gastroenterology**

Isolated Hepatic Polykystosis: About a Case in the Hépato-Gastroenterology Department of the National Hospital of Niamey

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Abstract

Polycystic liver disease is an autosomal dominant disorder, rare and benign in the majority of cases [1]. It is frequently associated with polycystic kidney disease, but can also be isolated in rare cases. Liver cysts result from abnormal growth of the biliary epithelium (cholangiocytes) or dilation of the peribiliary glands, due to the persistence of embryonic biliary structures. Isolated polycystic liver disease is generally asymptomatic, women are more affected than men and also have a greater number of cysts [2]. We report the case of a 40-year-old asymptomatic man with isolated polycystic liver disease Gigot III discovered during a health check-up. The clinical examination showed hepatomegaly without pain, of firm consistency, smooth surface, sharp lower edge with a hepatic arrow at 17cm. The kidneys are devoid of cysts, the liver function as well as the renal function are normal.

Keywords: Polycystic liver disease, cyst, autosomal dominant mode, asymptomatic, isolated.

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INTRODUCTION

Polycystic liver disease is a rare disease defined by the existence of multiple cysts within the liver parenchyma and/or the peribiliary glands. They are transmitted in an autosomal dominant mode. They are more frequent in women than in men (80%), especially after the fourth decade [1, 2]. PKH is most often associated with polycystic kidney disease, which determines the prognosis through the renal failure it can cause [4]. Most often asymptomatic, PKH can in rare cases be symptomatic; it may be complicated by infection or intracystic hemorrhage. Their management depends on the symptomatology, extent, distribution and anatomy of the cysts and may include percutaneous aspiration [2, 6] alcoholic sclerosis or fenestration of the cysts; a partial hepatectomy or even a liver transplant can be performed in rare cases where the hepatomegaly is disabling. A better understanding of the pathophysiological mechanisms of cyst formation has led to the proposal of somatostatin analogues in this indication, but the benefit of these treatments has not yet been clearly established [2, 3, 5]. However, most patients with isolated polycystic liver disease have a good prognosis and no treatment is needed. We report the case of isolated and asymptomatic polycystic liver disease in a 41-year-old man.

PATIENT AND COMPLIANCE

A 41-year-old patient came to consult for atypical cysts on ultrasound performed during an annual health check- up. The patient was asymptomatic, the physical examination showed hepatomegaly without pain, of firm consistency, smooth surface, sharp lower edge with a hepatic arrow at 17cm. Abdominal ultrasound concluded with atypical hepatic cysts without renal involvement, computed tomography showed polycystic liver disease (with multiple cysts disseminated throughout the liver) isolated without renal involvement, transaminases were normal, prothrombin level was unremarkable, the complete blood count was normal, the dosage of urea and serum creatinine was normal. Since there are no symptoms, no treatment was offered to the patient. Nevertheless, we invited him to do periodic checks and to comeback for consultation as soon as a symptom occurs, a family survey was also proposed but not carried out.

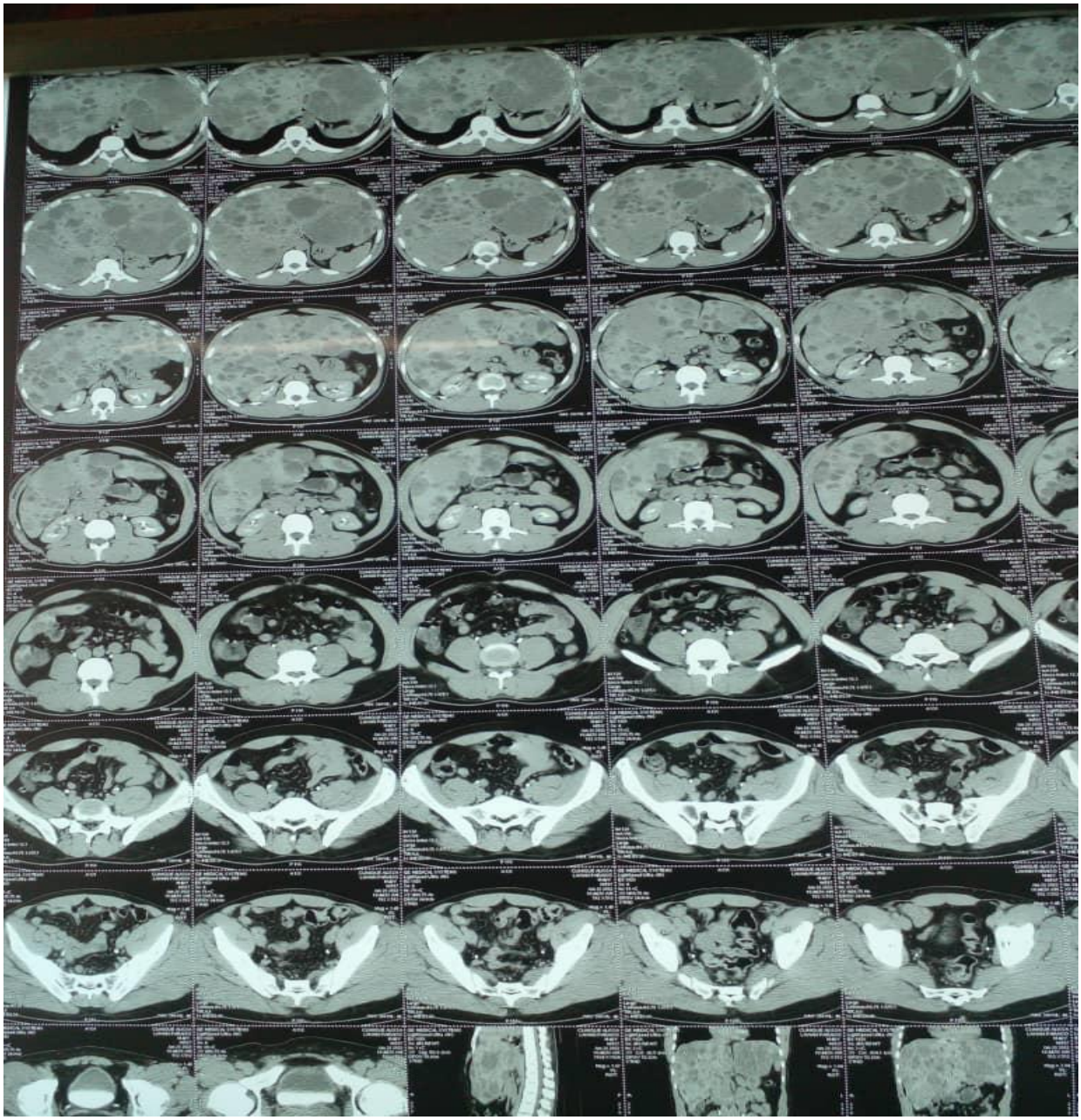

Figure 1: HEPATIC POLYKYSTosis

DISCUSSION

Our case is a 41-year-old man with no significant pathological history and no symptoms who was referred to us for cystic hepatic masses on abdominal ultrasound, which alone can make the diagnosis in the hands of a trained operator. Computed tomography showed isolated polycystic liver disease without kidney damage classified as Gigot III. This is a very rare case which is generally benign without any symptoms or complications, this is the observation made by most of the studies which stipulate that the prognosis is clearly linked to renal damage while the isolated liver damage is very rarely life-threatening [1, 2, 7]. In addition, larger and more numerous cysts are described in isolated polycystic liver disease than in hepatorenal polycystosis, on the other hand severe forms are more frequently observed in hepatorenal polycystosis. PKH is more common in women than in men [2]. Our case draws its particularity from the fact that it is a young man with isolated polycystic liver disease Gigot III and asymptomatic (Photo CT)... On the other hand, our case fits with the literature which stipulates that most polycystoses hepatic and even hepatorenal are asymptomatic and require no treatment, however periodic radiological monitoring is recommended.

CONCLUSION

Hepatorenal polycystosis is a benign and very rare pathology, isolated hepatic polycystosis (10%) [3,7] is even rarer. The latter is usually asymptomatic and only requires periodic monitoring. The limits of our

case were the impossibility of carrying out a family investigation given the "high cost of additional examinations

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