



EXCEPTIONAL CASE

Persistent pruritus as a rare and potentially serious manifestation of liver involvement in autosomal dominant polycystic kidney disease

Ana García-Prieto, Esther Torres, Andrés Hernández, Diego Barbieri and Almudena Vega

Nephrology Department, Hospital General Universitario Gregorio Marañón, Madrid, Spain

Correspondence and offprint requests to: Ana García-Prieto; E-mail: anamgarciaprieto@gmail.com

ABSTRACT

Polycystic liver disease (PCLD) is the most frequent extrarenal manifestation of autosomal dominant polycystic kidney disease (ADPKD), usually asymptomatic. We present a patient with ADPKD who developed cholestatic pruritus due to bile duct compression of multiple liver cysts. Because of severity of pruritus she received treatment with lanreotide to reduce liver volume and she was studied to be included in the liver transplant waiting list. She evolved favourably with medical treatment and she is now asymptomatic. In conclusion, persistent pruritus is a rare but potentially serious manifestation of PCLD, so it should be taken into account in patients with ADPKD.

Keywords: jaundice, lanreotide, polycystic kidney disease, polycystic liver disease, pruritus

BACKGROUND

Polycystic liver disease (PCLD) is the most frequent extrarenal manifestation of autosomal dominant polycystic kidney disease (ADPKD), which develops in up to 80% of diagnosed patients by the sixth decade [1]. Most patients remain asymptomatic and <20% develop liver disease. The main symptoms derive from hepatomegaly, which can lead to extrinsic compression of thoracic and abdominal organs [2]. Cholestatic pruritus is a rare complication of ADPKD that can even lead to liver transplant. We present the case of a patient with ADPKD who presented with persistent pruritus.

CASE REPORT

A 50-year-old woman diagnosed with ADPKD, undergoing home haemodialysis, presented with persistent pruritus. It started

2 months previously and had been worsening since then despite intensification of dialysis treatment and prescription of antihistaminic drugs, to the point of interfering with night rest. She referred to no abdominal pain, fever or recent modifications of her medication. Physical examination revealed mild jaundice and multiple scratch-related skin lesions. Liver function tests showed hyperbilirubinaemia (3.9 mg/dL) and normal aminotransferases but elevation of gamma-glutamyltransferase (1051 mg/dL) and alkaline phosphatase (792 mg/dL). Parameters related to the efficacy of dialysis treatment were within the normal range (creatinine 4.52 mg/dL, urea 45 mg/dL, potassium 4.1 mmol/L, phosphorus 3.8 mg/dL). The patient was admitted to the Hepatology department with diagnosis of obstructive jaundice. Abdominal computed tomography (CT) scan was performed, which showed multiple non-communicating cysts in liver and kidneys and mild dilation of intrahepatic biliary radicals in both lobes, attributable to extrinsic compression of

Received: 14.2.2018; Editorial decision: 30.5.2018

© The Author(s) 2018. Published by Oxford University Press on behalf of ERA-EDTA.

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com

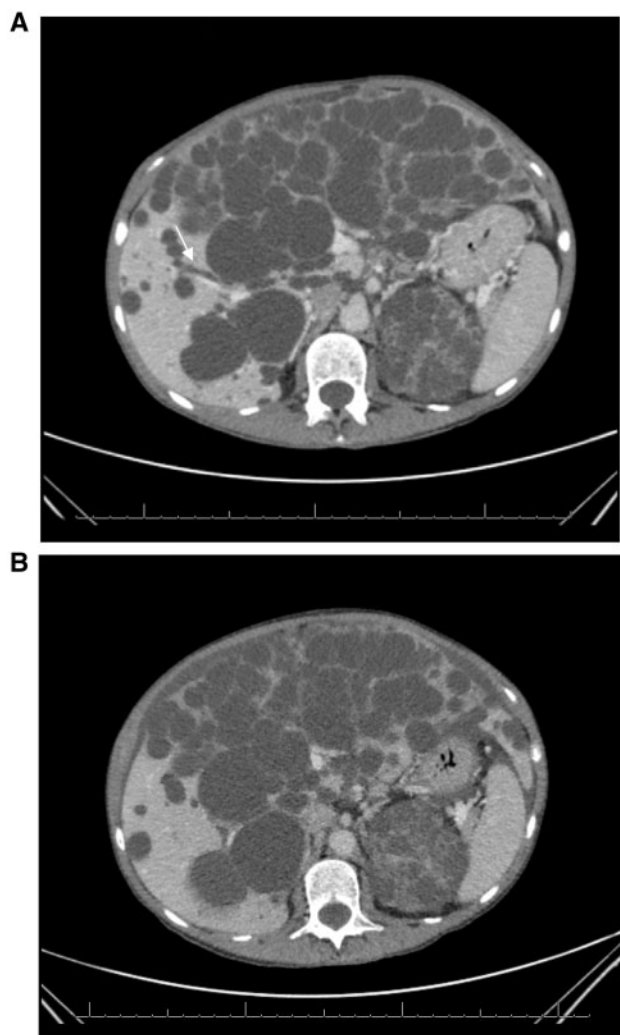


FIGURE 1: Abdominal CT scan before (A) and after (B) 6 months of treatment with lanreotide. Arrow in (A) shows mild dilation of intrahepatic biliary radicals due to extrinsic compression of multiple cysts that is not present after treatment, which led to reduction of cysts volume (B).

multiple cysts (Figure 1). Common hepatic duct and common bile duct appeared normal and thus not susceptible to drainage. Medical treatment of pruritus was intensified with cholestyramine and ursodeoxycholic acid but because of persistence of symptoms, lanreotide was started to reduce liver volume and the patient underwent multiple complementary tests to be included on the liver–kidney transplant waiting list. Symptoms improved 3 weeks after initiation of lanreotide and CT scan after 6 months of treatment showed decreased liver volume ($4023 \pm 120 \text{ cm}^3$ versus $3893 \pm 112 \text{ cm}^3$). She has since then been asymptomatic.

DISCUSSION

PCLD is an autosomal dominant disease usually associated with ADPKD. It is defined when >20 hepatic cysts are present and it represents the main extrarenal manifestation of ADPKD [1]. The

main risk factors for developing cysts are: age, sex (more frequent in young females), pregnancies, renal failure, oestrogen, hormone replacement therapy or oral contraceptives [3]. Our patient is a young woman with a long history of renal failure and two previous pregnancies, which could have contributed to the development and increase of liver cysts.

PCLD is asymptomatic in >80% of cases and diagnosis is often made incidentally. Symptoms usually derive from liver enlargement that can lead to extrinsic compression of thoracic and abdominal organs causing bloating, abdominal pain, gastroesophageal reflux, early satiety, nausea, vomiting, dyspnea, hernia, etc. Liver parenchyma is preserved despite cysts expansion, so liver failure is exceptional [2]. Presentation with pruritus alone due to bile duct obstruction is a rare manifestation, being described only in case reports.

Treatment of PCLD should be started when serious complications are present, as in the case of our patient. The main objective is to reduce symptoms by decreasing liver volume and options include invasive or medical measures. Percutaneous aspiration was not feasible in our case, because there was no large dominant cyst. Medical treatment using somatostatin analogues has shown effect in reducing liver volume by 6% in different clinical trials [4, 5]. They are well tolerated but seem to lose effectiveness after withdrawal. However, they have not been approved by the European Medicines Agency and can only be used in clinical trials or as compassionate use in severely symptomatic patients, like in our case. Nevertheless, the only curative treatment is liver transplant, but it is reserved for patients who do not respond to other treatment options. In our case, as the patient became asymptomatic after initiation of treatment with lanreotide, conservative management was chosen.

In conclusion, persistent pruritus is a rare but potentially serious manifestation of PCLD that should be taken into account in patients with ADPKD.

CONFLICT OF INTEREST STATEMENT

None declared.

REFERENCES

1. D'Agata ID, Jonas MM, Perez-Atayde AR et al. Combined cystic disease of the liver and kidney. *Semin Liver Dis* 1994; 14: 215–228
2. Ars E, Bernis C, Fraga G et al. On behalf of the Spanish Working Group on Inherited Kidney Disease. Spanish guidelines for the management of autosomal dominant polycystic kidney disease. *Nephrol Dial Transplant* 2014; 29: iv95–iv105
3. Gevers TJ, Drenth JP. Diagnosis and management of polycystic liver disease. *Nat Rev Gastroenterol Hepatol* 2013; 10: 101–118
4. Hogan MC, Masyuk TV, Page L et al. Somatostatin analog therapy for severe polycystic liver disease: results after 2 years. *Nephrol Dial Transplant* 2012; 27: 3532–3539
5. Neijenhuis MK, Gevers TJG, Nevens F et al. Somatostatin analogues improve health-related quality of life in polycystic liver disease: a pooled analysis of two randomised, placebo-controlled trials. *Aliment Pharmacol Ther* 2015; 42: 591–598