bilateral u obstrucción a la normal apertura y cierre de las válvulas.

El electrocardiograma puede mostrar alteraciones de las ondas T, que pueden ser picudas, negativas y simétricas, así como diversas arritmias, desde la extrasistolia ventricular aislada, hasta las taquicardias ventriculares y el bloqueo AV completo. El diagnóstico diferencial ecocardiográfico debe plantearse con fibrasomas, mixomas, rabdomiosarcomas o trombos intracardiacos. La RM es más sensible que la TAC en el diagnóstico de la hidatidosis cardíaca. El tratamiento en todos los casos debe consistir en la resección quirúrgica, acompañada de la administración muy prolongada, en ocasiones indefinida, de albendazol oral, salvo en algunos pacientes con quistes pequeños, calcificados, asintomáticos y con serología negativa. La recurrencia resulta excepcional y suele ser debida a la persistencia de quistes pequeños no detectados durante la cirugía.

Bibliografía


E. Alegria-Barrero a, b, D. Martín-Raymondi b, A. Alegria-Barrero y J. Barba
d
a Servicio de Cardiología, Hospital Universitario de Torrejón, Torrejón de Ardoz, Madrid, España
b Servicio de Cardiología, Hospital Santos Reyes, Aranda de Duero, Burgos, España
c Departamento de Cardiología, Hospital Universitario Madrid-Monterpríncipe, Madrid, España
d Departamento de Cardiología y Cirugía Cardiovascular, Clínica Universidad de Navarra, Pamplona, Navarra, España

a Autor para correspondencia.
Correo electrónico: ealegria@torrejonsalud.com (E. Alegria-Barrero).

http://dx.doi.org/10.1016/j.jce.2014.09.001

Polycystic kidney and liver disease: A diagnostic challenge

Enfermedad poliquística renal y hepática: un reto diagnóstico

The detection of single or multiple kidney cysts is very common, especially with advancing age, and it has no particular significance. On the other hand, there are patients who have multiple cysts, which, depending on a set of characteristics including age, the number of cysts and their distribution across several organs, may be included in the so-called polycystic diseases.

Polycystic liver disease is defined by the presence of 20 or more cysts in the liver. It may occur in the context of a rare disease such as autosomal dominant polycystic liver disease (ADPLD) or more often as a result of the autosomal dominant polycystic kidney disease (ADPKD). The prevalence of ADPLD is unknown due to the lack of population studies, and given its subclinical evolution it is rarely reported. In contrast, ADPKD is considered to be the most common hereditary nephropathy (prevalence 1/400–1/1000). Ninety five per cent of ADPKD cases are hereditary, with two known mutations: PKD1 (85%) – whose clinical manifestations are the earliest and more rapidly evolving – and PKD2 (15%). The renal manifestations of ADPKD usually involve increased kidney volume due to cortical cysts that alter the contour of the kidney, urinary tract and cyst infections. Liver involvement with multiple cysts is the most common extra-renal manifestation.

ADPLD, in turn, usually presents no renal involvement and its prognosis is benign. The genetic study of this disease is still limited. Two of the genes involved are known (PRKCSH and SEC63), but in approximately 80% of the cases genetic mutation is not identified. Extrahepatic (heart and intracranial) manifestations have been described, but in small studies. Despite the involvement of the organ, usually there is no change in the liver function.

Screening and diagnosis of these pathologies are performed based on imaging criteria according to age, family history and number of cysts in individuals (Table 1). Differential diagnosis between ADPKD and ADPLD is usually linear. We present a patient with polycystic kidney and liver disease with atypical clinical evolution whose diagnosis proved intriguing.

An 82-year-old woman went to the emergency department because of fever (38.5°C) and chills lasting for ten days, with diffuse abdominal pain. She was hemodynamically stable, and the abdominal palpation was painful on all sides, with no palpable masses. She had analytical elevated inflammatory and normal renal and hepatic function parameters and no changes in urinalysis. The urine culture revealed the presence of Klebsiella pneumoniae and Escherichia coli. A thoracic-abdominal-pelvic computerized tomography scan showed numerous renal and hepatic cysts.
Some cysts in the left kidney had heterogeneous content, suggesting infection. A diagnosis of acute urinary infection infectious of kidney cysts in the context of a possible ADPKD type 2 was advanced. The patient improved with antibiotherapy. Ultrasound testing performed to the patient’s daughters did not reveal any kidney or hepatic cysts disproportionate to their ages. The PKD2 mutation genetic study was negative. In the two-year follow-up period her renal function did not worsen and she had no other clinical manifestations of the disease.

Our patient showed multiple cysts meeting the ADPKD ultrasound diagnosis criteria, but is unusual due to her advanced age and the absence of repercussions on renal function and kidney’s size. Her kidneys had regular borders and multiple deep parapelic cysts, which are less common in ADPKD. The fact that the disease was not transmitted to any of the two daughters and the negativity of PKD2 gene mutation does not exclude the diagnosis, since the mutation detection is detected only in 60–70% of the cases as a consequence of the already known genetic and allelic heterogeneity of the disease.7

The possibility of ADPLD associated with the presence of parapelic cysts resulting from the advanced age of the patient was also considered. However, controversy remains since, according to diagnostic criteria, the large number of renal cysts points to a diagnosis of ADPKD.7 We found no justification for further genetic study, since PKD1 gene mutation would be highly unlikely, as the patient did not present rapid progression to kidney failure, while the ADPLD genetic study is still limited.

Thus, although one can usually make a simple linear differential diagnosis between ADPKD and ADPLD, there are some cases that pose relevant difficulties. Liver polycystic disease, expressed by the presence of over 20 cysts, is rare in ADPKD, since renal phenotype is dominant in the majority of patients; however, one subgroup present predominantly with hepatic cysts. Drenth et al. refer that a number of patients with ADPKD have kidney cysts and normal renal function, but extensive liver disease.8

### Bibliografía


M. Alves*, A. Miranda, M. Narciso, T. Fonseca

*Corresponding author.

E-mail address: marianaalves88@gmail.com (M. Alves).

http://dx.doi.org/10.1016/j.rce.2014.09.002