Case Study

Autosomal dominant polycystic kidney disease (ADPKD) with extra-renal manifestations misdiagnosed as liver hydatid cyst.

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Abstract

A 42 years old woman was presented at a private clinic with right renal colic and a history of liver hydatid cyst, abdominal examination revealed mild hepatomegaly, abdominal tenderness and palpable right kidney. The radiological findings demonstrated an enlarged liver and increased parenchyma echo-texture, well defined multiples cyst were seen at both lobes with different, size, thin wall regular outline, the largest one measuring 28.3 x 25.6 mm , with bilateral multiple renal cysts and was finally diagnosed as ADPKD with liver cyst as an extra-renal manifestation of ADPKD. This extra-renal manifestation of ADPKD as liver cyst may be misdiagnosed as liver hydatid cyst.

Keywords: ADPKD, liver cyst, renal cyst, polycystic liver disease and hepatomegaly.

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is described as multiple renal cysts with extrarenal cyst like liver cyst which is known as polycystic liver disease, the most common extrarenal manifestation of ADPKD with prevalence of 90% among ADPKD but it appears after renal cyst, with incidence of 0.2 % in general population [1].

ADPKD is most common in women at fourth-fifth decade of their life, the number and size of renal cysts is correlated with age, which ultimately leads to end-stage kidney disease [2].

Most of the patient with ADPKD stay asymptomatic and the diagnosis is usually incidental, however large liver cysts that are associated with ADPKD lead to hepatomegaly, abdominal distention and right loin pain [3].

The pathophysiology of ADPKD remains unknown, although it is related to the female steroid hormone levels, but that is more common in females [4].

Case presentation

A 42 years old woman was presented at a private clinic with right renal colic, nausea, vomiting, dysuria and malaise, on examination, vital signs were temperature 39°C, blood pressure 140/80 and heart rate 89 beats /min. Abdominal examination revealed mild hepatomegaly, abdominal tenderness and palpable right kidney while; other examinations were normal. Routine biochemical investigations showed followings results:

RBS=130mg/dl ,blood urea =39mg/dl ,serum creatinine =1mg/dl ,TSB =0.9 mg/dl , a white blood cell count was 13000 /cm2, 90% were neutrophils 8% lymphocytes 1% for eosinophils and basophils , ESR 90/first hr ,CRP =5.7mg/dl
and general urine examination reveals severe urinary tract infections with urate crystals and all liver function tests were normal.

She was treated with analgesic, diclofenac 100mg IM, antibiotic for UTI as cefotaxime 1g /12hrs and urinary antiseptics and then sent for abdominal ultrasound and MRI to explain the hepatomegaly and other intra-abdominal pathology.

The radiological findings demonstrated the following features: enlarged liver and increased parenchyma echo texture, well defined multiples cyst were seen at both lobes, vary in size, thin wall regular outline, the largest one measuring 28.3 x 25.6 mm, no pelvicalyceal system (PCS) dilatation, multiple crystals without visible stone, two simple renal cysts the largest one at mid pole measuring 45.3x45.4 mm.

Left kidney was normal in size, position and texture with smooth outlines, normal parenchymal thickness, no pelvicalyceal system (PCS) dilatation, multiple crystals without visible stone, renal cyst with multiples small cyst were seen the largest one measuring 34.1 x 32 mm. all other findings were normal, which gives a typical picture of autosomal dominant polycystic kidney disease ADPKD, figure (1 A, B, C).

Moreover, a full detailed history was undertaken that demonstrated the patient was previously diagnosed as a liver hydatid cyst and she was treated with Albendazole for three consecutive months without any improvement. We suggested her for an abdominal MRI, which revealed a picture of hepatorenal polycystic disease.

Positive family history for the same condition pointed out the final diagnosis of autosomal dominant polycystic kidney disease ADPKD with extra-renal manifestations.

After proper diagnosis she is now on a steroid injection (Sandostatin) 500mcg/ml sc/4 weeks with follow-up for any complications and drug side effects and according to the surgeon’s opinion she is currently unfit for surgical or laproscopic intervention due to huge liver cyst.
Discussion

The clinical presentation of patient was not corresponding and equivalent with the clinical feature of ADPKD with liver cyst, she was incidentally detected for the liver hydatid cyst, otherwise 70% of patients with liver cysts presented as painful hepatomegaly [5].

Regarding, the pathogenesis of hepatic cysts in ADPKD, a rise in the hepatocyte cAMP concentration escorts to alterations in apoptotic process and cholangiocyte proliferations [6].

Surgical interventions and hepato-renal transplantation are curative therapy but, this required talented surgeons and sophisticated devices which are unfortunately unavailable in most Iraqi hospitals, consequently, medical therapy are recommended mainly in large cysts or when patient is unfit for surgical interventions.

The pharmacotherapy for ADPKD with liver cysts is somatostatin analogue and rapamycin, octeriotide and lanoteride that are somatostatin analogue which act through inhibition of liver cAMP which is the critical factor in the pathogenesis of liver cyst in ADPKD, while rapamycin which is an immunosuppressive agent that demonstrated a controversy about the therapeutic effects on human and animal studies [7,8].

Extra-renal manifestations of ADPKD are rarely present in the liver as portal hypertension, ascites, esophageal variceal bleeding or Budd-chiari syndrome [9].

Figure (1-A, B and C): Ultrasonographic study shows multiple liver and kidney cysts.
In summary, the patient was initially misdiagnosed incidentally as liver hydatid cyst with simple renal cysts but finally she was managed by medical therapy due to large multiple cysts in liver and kidney and is on a steroid injection (Sandostatin) 500mcg/ml sc/4 weeks with follow-up for any complications and drug side effects.

Conclusion
A disease may have diverse symptoms but the need of an hour is to have proper diagnostic tools and talented medical professionals, so that the medical conditions may not be misdiagnosed and we may not lose the human lives.

Conflict of interest
The authors declare that there is no conflict of interest to reveal.

References