Case Report

UNILATERAL RENAL CYSTIC DISEASE (URCD)

Nessa A[1], Hossain AF[3], Mostafi M[2]

Abstract

Unilateral renal cystic disease (URCD) is a rare condition characterized by a unilateral enlarged kidney filled with multiple sized well-marginated cysts separated by parenchymal bands. Only a few cases have been reported in the literature till to date. It must be distinguished from other renal cystic diseases with which it shares radiological features. A case of young sailor diagnosed with URCD is reported here.

Introduction

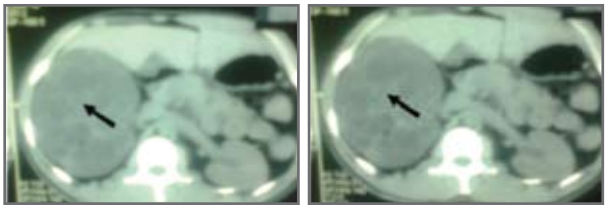
Unilateral renal cystic disease (URCD) is a rare and poorly understood condition first described in 1979[1]. Till 2010 only 55 cases have been reported throughout the world[2]. The disease is not familial and has also been known by other names like segmental cystic disease, localized cystic disease, multiple unilateral renal cysts and segmental polycystic renal disease[3]. URCD should be distinguished from other cystic entities such as autosomal dominant polycystic kidney disease (ADPKD), multilocular cystic renal nephroma, cystic dysplasia, and multiple renal cysts. A case of two developmental anomalies [URCD in the right kidney and Wolff Parkinson White (WPW) ECG pattern in a young adult sailor of Bangladesh Navy] is presented here.

Case Report

A 27 years old young Bangladeshi sailor presented to otolaryngologist with symptoms and signs of deviated nasal septum for which he was scheduled to undergo operative treatment (septoplasty). During routine pre-anaesthetic checkup, incidentally his blood pressure was found to be raised (170/106 mm of Hg). He denied any cardiovascular, respiratory, renal, endocrine or gastrointestinal symptoms. No family history of any cardiovascular or renal diseases was present. Subsequent physical examination revealed a palpable mass in the right flank of abdomen. No other congenital deformity was detected. Routine laboratory findings were unremarkable - namely blood picture, hemoglobin level (Hb%), serum urea, creatinine and electrolytes, urine analysis, protein profile, chest X-ray etc. Abdominal ultrasound revealed multiple cysts of varying sizes in the right kidney replacing normal renal tissue and normal left kidney (fig-1). Computed tomography (CT) scan revealed that right renal parenchyma is replaced by multiple varying sized cystic areas with unremarkable left kidney (fig-2a,2b) and there was no evidence of cystic disease in other organs or any urinary tract malformation. DTPA renogram revealed reduced perfusion in right kidney with split function of only 16.1% (left kidney-83.9%). DTPA GFR (glomerular filtration rate) was 8.9 ml/min in right kidney and 46.6 ml/min in left kidney (total GFR 55.5 ml/min), which is an indicator of early chronic kidney disease (CKD) (fig-3). His chest skiagram was normal. But electrocardiogram revealed WPW ECG pattern, although he was asymptomatic. Subsequent echocardiogram and exercise tolerance test were found to be normal. His parents underwent renal ultrasound and were unremarkable.

Fig-1: USG of KUB region showing multiple cysts in right kidney, replacing normal renal parenchyma and unremarkable left kidney.

1. Maj Azizun Nessa MBBS, FCPS, Armed Forces Medical Institute, Dhaka Cantonment; 2. Col Md Amzad Hossain Fakir MBBS, FCPS, FCPS, Nephrology (Pak), FRCP (Ireland) Classified Medical Specialist and Nephrologist, Combined Military Hospital, Dhaka Cantonment; 3. Col Mamun Mostafi MBBS, MACP (USA), MRACP(Australia), FCPS, FRCP (Ireland) ; Classified Medical Specialist and Nephrologist, Combined Military Hospital, Dhaka Cantonment.



(a) (b)

Fig-2(a, b): CT scan of abdomen (non contrast) showing multiple cyst in right kidney, replacing normal renal parenchyma and unremarkable left kidney, liver, spleen and other abdominal organs.

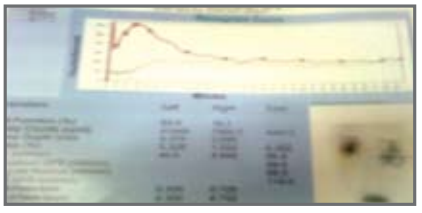


Fig-3: DTPA renogram with GFR revealed poorly perfused, mildly enlarged multicystic right kidney (GFR-8.9 ml/min as green line in graph) and normal functioning left kidney (GFR-46.6 ml/min as red line in graph).

So, the case was diagnosed as URCD of right kidney with early CKD, secondary hypertension and WPW ECG pattern. His blood pressure was controlled with Tablet Atenolol (50 mg once daily) and Tablet Amlodipine (5 mg once daily). Along with supportive treatment, he was advised for periodic follow up.

Discussion

Many different forms of cystic disease of the kidney exist ranging from simple cysts of no clinical significance to genetic abnormalities incompatible with life. The pathogenesis of URCD is unknown but acquired mal -developmental origin (congenital) is hypothesized, neither it is hereditary nor does it lead to progressive deterioration in renal function[4,5]. In view of the morphological similarity of the cystic change to the ADPKD, it is tempting to speculate that the pathogenesis of the cysts in URCD is the same[6]. Distinguishing this disease from ADPKD can be done by checking with the degree of confinement of the cystic disease. But its unilateral localization with an absence of cysts in other organs (like pancreas and liver), absent genetic background and absence of intervening normal parenchyma between cysts can distinguish well between URCD and ADPKD[2]. To diagnose URCD, characteristic CT findings are required in addition to genetic and clinical findings. URCD is characterized by cysts of varying sizes localizing in a diffusely enlarged kidney but not forming a distinct encapsulated mass and the absence of intervening normal parenchyma between the cysts unlike cystic renal nephroma[2]. Multiple cysts may be difficult to distinguish from URCD when confined to one kidney but they are less numerous than in URCD and they are predominantly located in the renal cortex whereas in URCD, they affect both cortex and medulla[7].

In unilateral dysplastic cystic kidney, the kidney is usually non-functioning as the collecting system is usually atretic or obstructed. Therefore in this condition the collecting system is usually not opacified on contrast enhanced imaging whereas in URCD the collecting system shows only a displacement[8,9]. Most patients of URCD are asymptomatic. Among those who are symptomatic, the most common symptoms include abdominal pain, haematuria (gross or microscopic), proteinuria and hypertension without impairment of renal function[10]. Reported patient presented with stage-II hypertension and he has mild impairment of renal function as evidenced by DTPA GFR (55.5 ml/min) and eGFR (88.8 ml/min), although his serum creatinine is normal (0.9 mg/dl).

In URCD, the contra lateral unaffected kidney in adult patients may occasionally show a few simple cysts as has been documented[7] and the evolution of unilateral disease into bilateral disease has been reported[2]. Moreover it is possible that development of complicated cysts due to rupture or infection of cysts, occurrence of malignancy or growth of a kidney may appear. Therefore these patients require long term follow up with functional imaging studies and surveillance[10].

Conclusion

The reported patient also have another developmental anomaly in his heart (by pass tract) causing WPW ECG pattern. So combination of two congenital anomalies in a person bears important clinical significance. It is a rare, benign, non surgical condition, which demands periodic follow up that have different treatment approaches and prognosis[11].

References

1. Cho KJ, Thornbury JR, Bernstein J, et al. Localized cystic disease of kidney. Angiographic-pathologic correlation. AJR. Am J Roetgenol 1979; 132: 891-895.

2. Yasunor M, Miho N, Kouichi H, et al. Non heriditary multiple cysts in unilateral kidney. NDT Plus 2011; 4: 73-74.

3. James BB, Attar K H, Rabbani S, et al. Secondary Polycythemia associated with unilateral renal cystic disease. Int Urol Nephrol 2007: 39: 933-58.

4. Par BS, Kim TH, Lim SJ, et al. Unilateral renal cystic disease. Korean J Urol 2007; 48: 652-54.

5. Bernstein J. A classification of renal cysts. In: Gardner KD Jr, Berstein J, editors. The cystic kidney, Dordrecht. The Netherlands: Kluwer; 1990.p.147-170.

6. Lin SP, Chang JM, Chen HC, et al. Unilateral renal cystic disease, report of one case and review of literature. Clin Nephrol 2002; 57 : 320-324.

7. Hwang Dy, Ahn C, Lee JG, et al. Unilateral renal cystic disease in adults. Nephrol Dial Trasplant 1999; 14: 1999-2003.

8. Lec S, Park SK, Kang SK, et al. A case of unilateral renal cystic disease. Nephrology 2004; 9: 31-32.

9. Bisceglia M, Galliani CA, Senger E, et al. Renal cystic disease, a review: Adv Anat Pathol 2006; 13: 25-26.

10. Venkata S, Katabathina MD, Goppi Kota MD, et al. Adult renal cystic disease: A genetic, biological and developmental process. Radiographics 2010 October; 30: 1509-1523.

11. Farinha A, Assancao J, Vinhas J. Unilateral renal cystic disease, report of an additional case of a rare disease. J Nephrol Hypert 2011; 25 (2): 163-165.