UNILATERAL CYSTIC RENAL DISEASE (URCD)

Nessa A1, Hossain AF2, Mostafi M3

Abstract

Unilateral cystic renal disease (URCD) is a rare condition characterized by a unilateral enlarged kidney filled with multiple sized well-defined cysts separated by parenchymal bands. Only a few cases have been reported in the literature till 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 1979.3 Till 2010 only 55 cases have been reported throughout the world.4 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. URCD should be distinguished from other cystic entities such as autosomal dominant polycystic kidney disease (ADPKD), multilocular cystic renal fibromata, 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 Bangladesh sailor presented to otolaryngologist with symptoms and signs of deviated nasal septum for which he was scheduled to undergo operative treatment (septotomy). During routine pre-anesthetic checkup, incidentally his blood pressure was found to be raised (170/96 mm of Hg). He denied any cardiovascular, respiratory, renal, endocrine or gastrointestinal symptoms. No family history of any cardiovascular or renal disease was present. Subsequent physical examination revealed a palpable mass in the right flank of abdomen. No other congenital anomaly was detected. Routine laboratory findings were unremarkable—namely blood picture, hemoglobin level (10%), serum urea, creatinine, and electrolytes, urine analysis, protein profile, chest X-ray etc. Abdominal ultrasound revealed multiple cystic spaces of varying size 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). His chart skiascope 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. 2a-b: DTPA images with GFR revealed grade I rarely patient, mildly enlarged multicystic right kidney (GFR 8.9 ml/min as green dot) and normal normal functioning left kidney (GFR 46.6 ml/min as red dot) in graph.

So, the case was diagnosed as URCD of right kidney with early CKD, second hyperplasia 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 is hereditary nor does it lead to progressive deterioration in renal function4. 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.5 Differentiating this disease from ADPKD can be done by being vigilant during the early stages of the condition 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. 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 neoplasms. 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.

In unilateral dysplastic cystic kidney, the kidney is usually non-functioning as the collecting system is usually stilet or obstructed. Therefore in this condition the collecting system is usually not opacified on contrast enhanced imaging whereas in URCD the collecting system shows it is only a displacement.6 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 function7. 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 (0.5 mg/dl).

In URCD, the contralateral unaffected kidney in adult patients may occasionally show a few simple cysts as has been documented2 and the evolution of unilateral disease to bilateral disease has been reported. Moreover it is possible that development of complicated cysts due to rupture or infecion of cysts, delayed malignancy or growth of a kidney may appear. Therefore these patients require long term follow up with functional imaging studies and surveillance8.

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.1 It is a rare, benign, non-surgical condition, which demands periodic follow up that have different treatment approaches and prognosis.

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Case Report

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Abstract

Unilateral renal cystic disease (URCD) is a rare condition characterized by a unilateral enlarged kidney filled with multiple sized water containing cysts separated by parenchymal bands. Only a few cases have been reported in the literature till 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 1979.1-7 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), multicystic dysplastic kidney, 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.

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A 27 years old young Bangladeshi sailor presented to otolaryngology with symptoms and signs of deviated nasal septum for which he was scheduled to undergo operative treatment (septoplasty). During routine pre-anesthetic checkup, incidentally his blood pressure was found to be raised (170/96 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 or neoplastic abnormality was detected. Routine laboratory findings were unremarkable – namely blood picture, hemoglobin level (10%), serum urea, creatinine, and electrolytes, urine analysis, protein profile, chest X-ray etc. Abdominal ultrasound revealed multiple water containing 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 8.5 ml/min in right kidney and 46.5 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 x-ray 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.

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 maldevelopmental origin (congenital) is hypothesized, neither it is hereditary nor does it lead to progressive deterioration in renal function2. In view of the morphological similarity of the cystic changes to the ADPKD, it is tempting to speculate that the pathogenesis of the cysts in URCD is the same.2 Distinguishing this disease from ADPKD can be done by being 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. 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 neoplasms.3 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.4 In unilateral dysplastic cystic kidney, the kidney is usually non-functioning as the collecting system is usually stenotic 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 slight dilatation.5 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.6 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 (38.8 ml/min), although his serum creatinine is normal (0.5 mg/dl).

In URCD, the solitary uninfected kidney in adult patients may occasionally show a few simple cysts as has been documented7 and the evolution of unilateral disease to bilateral disease has been reported. Moreover it is possible that development of complicated cysts due to rupture or infecion of cysts (by past malignancy or growth of a kidney may appear. Therefore these patients require long term follow up with functional imaging studies and surveillance.7

Conclusion

The reported patient also have another developmental anomaly in his heart (by past trauma) 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.

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References

CASE REPORT ON PSEUDOCYSEIS
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Abstract
Introduction: Rare condition in which a nonpregnant patient has the signs and symptoms of pregnancy, such as abdominal distention, breast enlargement, pigmentation, cessation of menses, and morning sickness1. Pseudocyesis is defined as the conviction of a non pregnant woman that she is pregnant, occurring with symptoms associated with pregnancy. It excludes delusions of pregnancy during psychosis, false pregnancy in malingering, endocrine disorders such as the galactorrhea-amenorrhea syndrome, and pelvic or abdominal tumours causing symptoms of pregnancy. Pseudocyesis is found especially in societies where there is much cultural pressure on women to have children. It may be considered as a defence against the wish for pregnancy, fear of pregnancy, or even resolving conflict between the two. Its origins have usually been traced to a disorder of personality. Chronic social deprivation and problematic relatives ships figure prominently. Several authors mention the naivety, gullibility, and lack of sophistication of these patients1.

A great number of terms are given to this state: Pseudocyesis, spurious pregnancy, phantom pregnancy, imaginary pregnancy, hysterical pregnancy, and simulated pregnancy. Although the term "false pregnancy" is frequently used synonymously in reference to this condition, it really is a misnomer. "False pregnancy" should denote only such a condition in which malingering or intentional deception is attempted.

Pseudocyesis is most commonly found in the neuotic and less intelligent types of individuals, especially those suffering from mental and emotional changes. However, occasionally it may fool even an intelligent woman who has had previous pregnancies.

Case report
A 52 years old recently divorced multipara women bailing from rural background of Bangladesh, referred by gynaec & Obstetrics outpatient department was admitted in Psychiatry dept of BSMMU with the symptoms of scanty fluidy vaginal discharge in each month for last 3 months, before that she experienced of amenorrhea for 3 months. She had morning sickness, nausea and occasional vomiting, fetal movement, abdominal distortion for last 10 months, breast milk secretion for last 6 months. At eighth month from onset of amenorrhea she developed labour pain which she claimed to be similar to previous labour pain. Till last interview patient believed herself as pregnant. All previously mentioned pregnancy related symptoms persisted despite leaving no positive findings in her investigations. Even though she repeatedly requested for caesarean section. None of his family members or relatives suffered from such or other psychiatric illness. She had 63 step mothers & 9 step siblings. She was brought up by none of her step mothers whereas she had good relationship with all step mothers as well as step siblings. She got married first time in 1999, had 4 children & ultimately was divorced. Then she fell in love affair and got into second marriage in 2005 and it was not accepted by both families. They