Late Presenting Unilateral Multicystic Dysplastic Kidney with Ipsilateral Pelviureteric Junction Obstruction and Hypertension in an Adolescent: An Incidental Finding

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ABSTRACT

15 year old female adolescent who presented on account of recurring abdominal pain of 4 years. Nil preceding history of trauma or fall. Not a known sickle cell disease patient. Nil previous abdominal surgery. Blood pressure at presentation was elevated for age and gender. Investigations revealed a non functioning dysplastic left kidney with hydronephrosis. She subsequently had left nephrectomy. Blood pressure thereafter normalised. She remains clinically stable after 12 months of follow up.

Key words: Abdominal pain, Hypertension, Multicystic dysplastic kidney, Congenital anomalies of the kidney and urinary tract, Pelviureteric junction obstruction

Aim: To highlight the importance of detailed clinical clerkship, thorough physical examination, high index of suspicion and exhaustive relevant investigations (laboratory and radiological) for all cases presenting to any healthcare facility.

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Presentation

15 year old senior secondary student who resides with her parents in Southwest Nigeria. Presented on account of recurring abdominal pain of 4 years' duration. Onset of pain was insidious. Nil history of trauma or fall prior to onset of symptom. Nil history of frequent passage of loose stool or vomiting. Nil associated fever. No history of reduced urination. Nil urinary frequency, urgency or urge incontinence. Nil history of bedwetting. Pain is generalised and is not referred to other parts of the body. Aggravated by exertion. Relieved by the use of analgesic and rest. Pain occurs about once in three months, intermittent within a 24-48 hour period and then resolves. It is not associated with her menstrual cycle.

Nil previous history of hospital admission. Nil prior blood transfusion or surgery. Not a known HbSS patient (Hb Genotype AA). Nil known drug allergies. Attained menarche at the age of 11 years. Menstrual flow lasts 4days in a 28-day cycle. Nil history of coitarche or contraceptive use. No dysmenorrhea, intermenstrual bleeding or mucopurulent vaginal discharge. Product of term, uneventful gestation. Details of the prenatal ultrasound findings could not be recalled by the mother. Neonatal period was not adversely eventful. She has no known food or drug allergies. She is in the upper 10th percentile of her year in school. The 3rd child in a monogamous family of three children. Nil family history of renal disorders. There was no history of parental consanguinity.

Examination revealed a healthy-looking female adolescent who was not in any obvious respiratory or painful distress. She was neither pale nor icteric. Breasts, axillary hair and pubic hair were all at Tanner Stage 5 of development.

Anthropometric Findings:

Weight: 72.5kg

Height: 1.54m

Body surface area: $1.76m^2$

Abdomen was full and moved with respiration. There were no areas of tenderness. Intra-abdominal organs were not palpably enlarged. Bowel sounds were normal. Digital rectal examination did not reveal any abnormality. Cardiovascular system examination revealed a blood pressure of 134/85mmHg (which was elevated for her age and gender). Other cardiovascular findings were normal. The other systemic examination findings were normal.

The initial working diagnoses were:

- ?Functional Abdominal Pain in a female adolescent
- Secondary Hypertension ?cause in an adolescent

Table 1: Initial investigation findings

FBC	E,U,Cr	Abdominal USS	CT Abdomen	Others
HCT: 34%	K^+: 3.5mmol/L	A huge multiloculated cystic mass (measuring 232x133mm) with homogenous internal echoes was visualised in the left renal bed. No normal kidney was visualized, on the left.	Bilateral obstructive uropathy and non functioning left kidney 2^o to ?PUJ Obstruction.	Renal scintigraphy (MAG3) showed a non-functioning left kidney with no obstruction on the right.
WBC: 6,490/µL	Na^+: 127mmol/L			Urinalysis: Normal findings

Neutrophil: 61%	Cl^-: 117mmol/L			
Lympocytes: 35%	HCO_3^-: 20mmol/L			RVS: Non reactive to HIV I & II
Eosinophils: 1%	Urea: 9mg/dL			
Basophils: 2%	Creatinine: 0.7mg/dL			HBsAg: -ve
Monocytes: 2%				
Platelets: 263,000/µL				AntiHCV: -ve

MAG3: Mercaptoacetyltriglycine

Treatment

Parents and the patient were informed about the investigation results. They were also counselled on the need for left nephrectomy for long term optimal blood pressure control in order to prevent complications associated with persistently elevated blood pressure. She was commenced on low dose antihypertensive (oral Amlodipine 2.5mg daily) which she had for a week and was scheduled for surgery. Blood pressure prior surgery was 129/78mmHg.

Intra operative findings were:

- Grossly normal right kidney; no compensatory hypertrophy
- Markedly dilated multi lobulated left kidney of about 30cm by 20cm extending towards the midline from the left lumbar region
- Dilated renal pelvis with constriction at the left pelviureteric junction
- Normal bowel, spleen and liver

Course

Immediate post-operative period was essentially not adversely eventful. Blood pressure was initially elevated (range 145/80 – 150/85mmHg) but it subsequently normalised over the succeeding 24hour period (120/70 – 126/72mmHg). She had adequate analgesia for pain relief. Post-operative haematocrit was 33%. Graded oral sips were commenced on post-operative day (POD) 3 and she was tolerating full enteral feeds by POD4. She was allowed home on the sixth post-operative day (POD6). Anti-hypertensive was discontinued after the first week of follow up. Blood pressure readings remained optimal. The follow up periods were as follows: one week, six weeks, three months, six months and twelve months post surgery.

Table 2: Anthropometry, Blood Pressure, Serial Serum Electrolytes, Urea and Creatinine in the immediate post-operative period and during follow up

Parameter	**24hours**	**1 week**	**6 Weeks**	**3 Months**	**6 Months**	**12 Months**
Weight (kg)	-	72.5	77.0	73.5	80.5	78.0
Height (m)	-	1.54	1.55	1.55	1.55	1.55
Body surface area (m^2)	-	1.76	1.82	1.78	1.86	1.83
Blood pressure (mmHg)	122/70	120/75	118/70	120/71	116/70	120/72
K^+ (mmol/L)	4.2	3.4	3.9	4.3	4.0	4.0
Na^+ (mmol/L)	116	140	126	137	137	133
Cl^- (mmol/L)	122	114	115	115	107	111
HCO_3^- (mmol/L)	21	29	19	22	22	19
Urea (mg/dL)	5	7	7	9	11	6
Creatinine (mg/dL)	0.6	0.5	0.6	0.8	0.7	0.7

Summary of histopathologic finding of the specimen (Left kidney)

- A nephrectomy specimen with overlying perinephric fat
- Weighed 500g

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- Dimensions: 12cm x 7cm x 6cm
- Cut section shows replacement of normal renal tissue by multiple large cystic cavities lined by urothelium in some areas and flattened epithelial cells in other areas
- There was extensive fibrosis of the interstitium
- Overall features were in keeping with multicystic renal dysplasia

The other first degree relatives were evaluated for hypertension and were also screened for the possibility of kidney dysplasia with ultrasound sonography. The outcomes were not suggestive of kidney disease.

DISCUSSION

Multicystic dysplastic kidney (MCDK) is one of the common renal cystic diseases that are identified as

congenital anomalies of the kidney and urinary tract (CAKUT).[1] Other examples of CAKUT include the following: Polycystic kidney disease (PKD), Duplex kidneys, Horse shoe kidneys, Ectopic kidneys and Ectopic ureters.[2,3] Urinary malformations in patients with MCDK consist of vesicoureteral reflux (VUR), ureteropelvic junction obstruction, and kidney stones.[2] They are the commonest form of congenital anomalies in the paediatric age group.[4] The genetic causes for most cases remain unknown.[5,6]

Although the pathogenesis of MCDK remains unclear, failure of the ureteric bud to integrate and branch appropriately into the metanephros during development in early childhood is one of the postulates.[2,3,7] The incidence of MCDK varies widely, and it has been reported to be more frequently unilateral, with a higher prevalence in the left kidney,[4,8] although some other studies have a higher prevalence on the right.[9,10] Some studies put the estimate at in 1 in every 1000 to 4300 live births.[7,11]

Majority of cases are detected either during routine antenatal ultrasonography studies or shortly after birth.[3,4] In extremely rare cases, they were not detected until adolescence or in early adulthood.[10,11] The child reported in this case study was one of the late presenters. Reasons for the late presentation could be due to: ease of access to over-the-counter medications in our environment (for pain relief),[12,13] apparent clinical stability of the patient (symptom not severe enough to necessitate hospital admission) and the level of clinical expertise at the health facilities she had previously presented to before arrival at our facility.

Pain (abdominal and flank), painful micturition and poor urinary stream have been identified as some of the presenting symptoms in children with CAKUT with chronic abdominal pain either being silent or vague.[14] MCDK, however, is generally not symptomatic.[15] CAKUT have been reported to be the leading cause of chronic renal failure.[2,4] The frequency of hypertension in children with unilateral MCDK (UMCDK) was reported as 5.4 per 1000 in a systematic review and those with an abnormality in the opposite kidney had a higher risk of hypertension.[16] Two mechanisms have been postulated for the pathogenesis of hypertension in UMCDK. Primarily, hyperreninemia due to the dysplastic kidney and secondarily, hyperfiltration in the contralateral kidney.[1,17] Older studies state that following the removal of a unilateral poorly or nonfunctioning kidney (including MCDK), resolution of hypertension occurs,[17] however, more recent studies favour a conservative approach.[9,10] The patient in this study was an exception because of her late presentation, hence, surgical removal of the non-functioning left kidney was opted for because she had grown well past the age when spontaneous involution of the dysplastic kidney was expected to have occurred.[8,10,18] Removal of the dysplastic kidney reduces excessive renin production which contributes to hypertension in UMCDK.[1,17] She has been followed up for twelve months since she had left nephrectomy, and there has been no record of hypertension during any of the outpatient visits.

Malformations of the reproductive system on the ipsilateral (same) side and various other malformation syndromes are often associated with CAKUT.[3,4] Examples include seminal vesicle cyst (in males), Müllerian anomalies and Gartner's cyst (in females).[19,20] The radiological investigations done on this patient did not suggest any reproductive organ abnormality and the adolescent is being kept on active surveillance by the gynaecological team of the study centre. No other malformation of any organ or system was detected in patient of this case study. Studies have reported that about 10-50% of children with CAKUT have affected family members (first degree relatives) with a positive history of renal anomalies or disorders of the urinary tract (e.g Renal agenesis, Nephrolithiasis, Pelvic Kidney, MCKD), ,[8,21] however, the relatives of the child in this case study were not

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affected. Because the initial screening done on the relatives was negative for CAKUT, genetic evaluation is not being considered for them. However for the child, it is being actively considered to predict the impact of the disease on her reproductive capacity.

This study is of note because of the following reasons. First, most cases of MCDK are diagnosed prenatally and involution of the affected kidney (either complete or partial) is expected to have occurred latest by the fifth year of life.[8,10,18] Second, the patient presented with recurring abdominal pain of four years (with onset in her teenage years) without a definite aetiology of the pain being arrived at until presentation at our facility. Vital signs at presentation in our facility showed elevated blood pressure for age and gender. This necessitated further investigations and eventual nephrectomy. If the diagnosis of the multicystic dysplastic kidney had been missed, hypertension would have persisted which places the patient at a significantly higher risk of chronic renal failure compared to the general population.[22] This underscores the importance of detailed clinical clerkship, thorough physical examination, exhaustive relevant investigations (laboratory and radiological) for all cases presenting to any healthcare facility in order to arrive at definite diagnosis and offer the most appropriate modality of care.

CONCLUSION

Unilateral multicystic dysplastic kidney, although usually diagnosed very early, occasionally presents late and systemic hypertension in an older child or adolescent may be a clue to this diagnosis.

Strengths of the Study

Apart from the radiologic and nuclide studies, we were able to get a histological confirmation which is the hallmark of the diagnosis of renal dysplasia.

LIMITATION

The patient has only been followed up for a 12-month period. A longer duration of follow up would have been more appropriate in determining the long term outcome of the patient vis-à-vis kidney function and reproductive capacity.

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Disclosures

The authors have no conflicts of interest to declare.

Consent

Written informed consent from the parents (and assent from the patient) was obtained.

Ethical Clearance

The medical ethics committee of the study centre granted a full waiver for a retrospective examination of the patient's records for the purpose of this case study.

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