CASE REPORT

UNILATERAL KIDNEY DUE TO TERATOGENIC EVENTS IN AN 18YEAR OLD ABSENT

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ABSTRACT

Renal absence, unilateral or bilateral, may result from progressive renal involution due to teratogenic effects. We report a case of an 18-year-old female who presented with unilateral renal absence after being involved in an RTA with the only complains from her being lower abdominal pains. The management of unilateral renal absence is always good provided the solitary kidney is healthy and the prognosis is good.

Key words: Renal agenesis, Multicystic dysplastic kidney, Renal aplasia.

INTRODUCTION

Renal absence is the non-existence of a kidney in a human which can be due to either renal agenesis or nephrectomy (which is done due to an anatomical anomaly, tumour, following a severe traumatic injury or a result of kidney donation). [1]

Renal agenesis can be unilateral or bilateral. When the fetal kidney fails to develop, oligohydramnios ensues and this is immediately followed by pulmonary hypoplasia, which might result to perinatal mortality. This sequence of events is more severe in cases with bilateral renal agenesis. [2] Studies have shown that there is a genetic basis for the occurrence of renal agenesis. [3]

It is interesting to note that the mother of the patient had gestational diabetes mellitus while pregnant with her. The patient's family however, believe that the road traffic accident she was involved in was the cause of the unilateral renal absence.

CASE REPORT

O.L is an 18-year-old female who presented at the Obafemi Awolowo University Teaching Hospital complex, Ile-Ife, Osun State, on account of right sided lower abdominal pains and tenderness of 16 months duration.

Patient was apparently healthy until 16 months prior to presentation when she had a road traffic accident that involved a motorbike. She sustained a wound at the right lumbar region posteriorly and presently has a scar at that region. She was hospitalized for about 2 weeks in a Missionary medical centre in Ile-Ife and before being discharged she was informed that the accident had affected her right kidney.

She started having right lower abdominal pains a week after discharge. The pains was insidious in onset, and moderate in intensity. The pain was intermittent and when it commenced, it usually progressed from slight discomfort then through mild to

moderately intense pains before disappearing. The duration of the episode of the pains was within 10 minutes and when it occurred, she had to terminate whatever she was doing and remain still. However, a month prior to presentation the pains became severe in intensity with increased frequency and duration. She always had to lie down flat for a while so that the pains will subside. She claimed to have never taken any analgesic for pain relief.

No triggering factor had been noted and there was no history of radiation of pains to the groin. There was no associated nausea, fever or hematuria. There was no history of abdominal distension, dysuria or difficulty to pass urine after the road traffic accident.

Patient's mother had gestational diabetes while pregnant with her. However, the labour was uneventful and patient was active after delivery.

On examination at presentation, she was healthy-looking, not pale, not feverish, anicteric, and not in any form of distress. Her blood pressure was 110/60mmHg. Patient's abdomen was scaphoid. No ballotable organs were palpated, however, mild to moderate right iliac fossa tenderness was elicited. Tympanitic sounds were present on percussion.

An abdominopelvic ultrasound scan was requested (Fig. 1 and 2) which showed that her right kidney was not at the renal bed and was not present at the right pelvic region.

An intravenous urography (IVU) was done (Fig. 2 and 3) and only the left kidney excreted the administered contrast agent. There was no ectopic kidney in the chest region.

An impression of unilateral renal absence was made secondary to renal agenesis. However, patient's mother claimed that it was the accident that damaged her right kidney. An abdominopelvic ultrasound scan report done 6 months prior to presentation at a private diagnostic centre stated that both kidneys were present and normal but she had also been told 16 months ago that the accident had affected her right kidney.

Blood chemistry analysis, lipid profile, Serum protein, and urinalysis were all within normal limits. Patient was booked for a follow-up in 6 months' time where her kidney function status and blood pressure will be assessed. She still complains of mild non-specific abdominal pains.

DISCUSSION

Unilateral renal absence due to agenesis has an incidence of 1 in 1500 births, with a male to female ratio of 1.8 to 1. It occurs frequently on the left side. In the index case, it was on the right side. Normal renal development depends upon a normal ureteric bud, which undergoes orderly branching and penetrates the metanephric blasterma at about the 5th week of gestation. When there is failure of the ureteric bud to form or absence of the nephrogenic ridge, the kidney does not develop normally. [2,4,5]

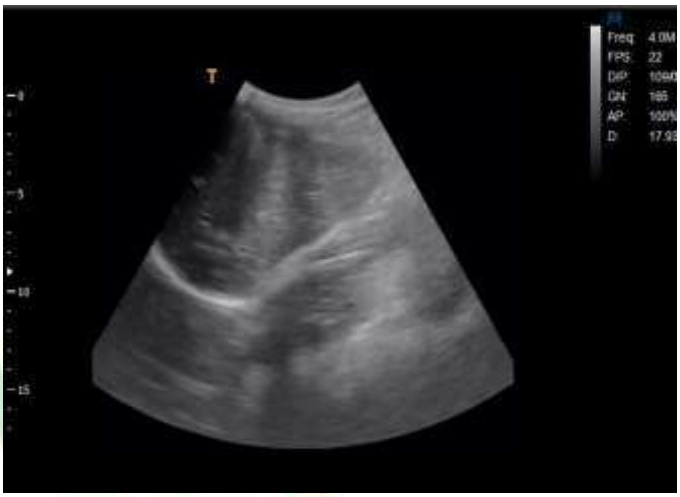

FIG. 1: Longitudinal ultrasound scan of the abdomen shows that the right kidney is not at the renal bed.

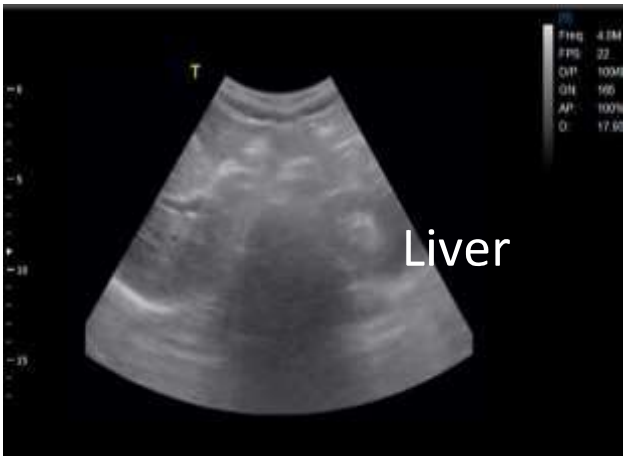

FIG. 2: Transverse ultrasound scan of the abdomen shows the presence of the left kidney but the right kidney is not present at its bed (blue star).

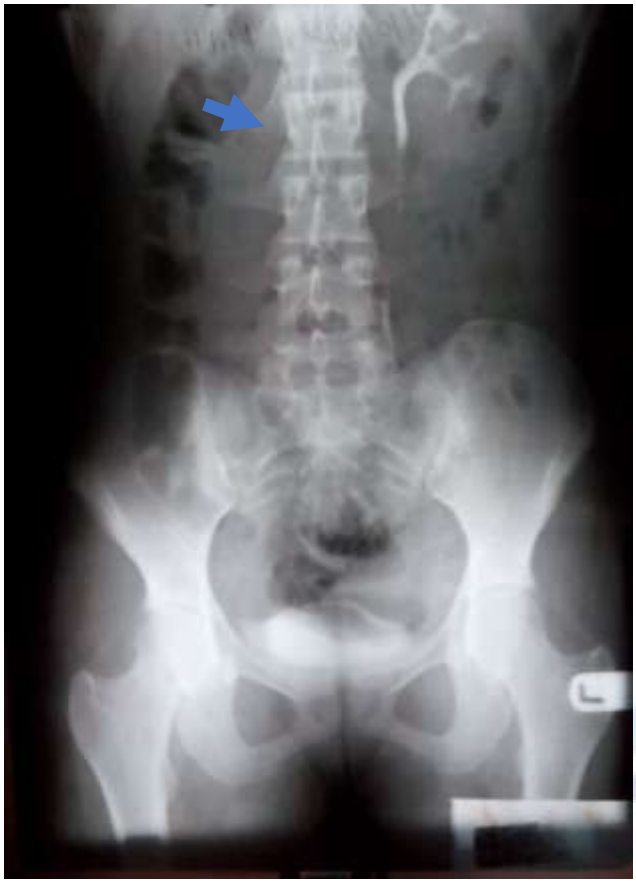

FIG. 3:
Intravenous urography. A 10minutes radiograph which shows only the left kidney excreting contrast medium into the urinary bladder. Normal bowel gas pattern is seen outlining the ascending colon (blue arrow).

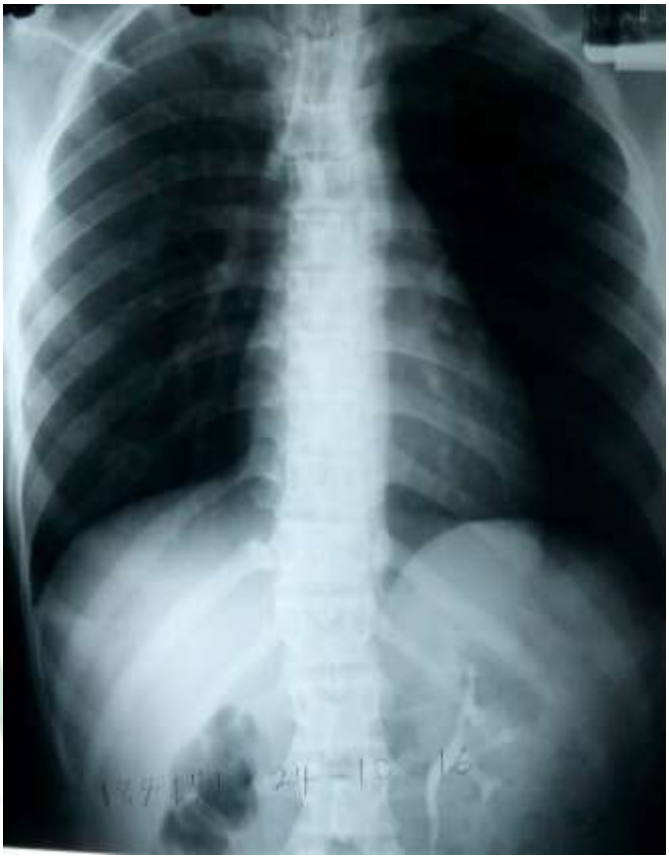

FIG. 4:
Intravenous urography. A 15minutes radiograph showing the absence of a functional ectopic right kidney in the chest. The pelvi-calyceal system of the left kidney is seen at the left renal bed.

Renal agenesis could also be due to the absence of transcription factor WT1 that influences growth factor FGF-2 and BMP-7 to prevent apoptosis of metanephric cells. [2] Attempts to discover the genes responsible for agenesis, hypoplasia and dysplasia have been challenging. [6] Unilateral renal agenesis is believed to be inherited as an X-linked dominant trait and also associated with Kallmann's syndrome. [2]

Autopsy studies have shown that renal agenesis can occur with the development of either a partial or a completely normal ureter, and a rudimentary kidney can be present in some cases where there is no

identifiable ureter. Mullerian duct abnormalities occur frequently in unilateral renal absence such as unicornuate uterus and uterus didelphys. [4,5] The patient's uterus and ovaries were sonographically normal.

Associated genital anomalies are 3 to 4 times more frequent in females than in males. The condition can be associated with Mayer-Rokitansky-Kuster-Hauser syndrome, in which there is congenital absence of the uterus and vagina. In the male, absence of seminal vesicle, vas deferens or the epididymis unilaterally may occur. [4] Absence of the vas deferens can be bilateral even in unilateral renal agenesis. [7] Renal agenesis is closely associated with ipsilateral congenital anomalies of urinary, cardiac and skeletal systems [2] and thus associated with VACTERL syndrome. [4]

The adaptive process culminates in hypertrophy and hyperfiltration of the nephrons in the solitary kidney which in turn produces lesions of the glomerular basal membrane. This leads to proteinuria (albuminuria), hypertrophy of the solitary kidney and focal segmental glomerulosclerosis later in life. [10-12] Vesico-ureteric reflux is the most common abnormality in the solitary kidney with persistent ipsilateral flank pains due to recurrent urinary tract infections. Hypertension and mild renal insufficiency are also well documented findings in individuals affected with unilateral renal agenesis. [2,4,10] Non-specific lower abdominal pains may also be a clinical presentation. However, most times it is asymptomatic and detected accidentally. [2,4] The patient's main complaint was lower abdominal pains.

Multicystic dysplastic kidney involute before birth, or in the 1st year of life, due to apoptosis to a rudimentary non-appreciable form and this is responsible for an increase in the incidence of renal agenesis. [1,4] Some researchers are of the opinion that multicystic dysplastic kidney's degeneration to renal aplasia, after birth, and subsequently to renal agenesis is probably accelerated by trauma. [7,8] Szmigielski et al [9] found that pre-existing renal abnormalities predispose the kidneys to an increased risk of injury and a lower probability of renal salvage following blunt abdominal trauma. Therefore, trauma to an abnormal kidney such as an aplastic kidney which may rapidly lead to involution might progress to renal agenesis. This could have been the explanation of how the road traffic accident (RTA) led to the disappearance of the right kidney in the index case. Based on the theory of the patient's mother, probably the right kidney was already aplastic.

The observation from the study embarked upon by Szmigielski et al [9] on renal trauma cases was that there was no incidence of renal disappearance following trauma to an erstwhile normal kidney. Many researchers believe that renal agenesis, renal aplasia, and multicystic dysplastic kidney may be genetically related, at least in some, and can be viewed as a continuum, since these anomalies are found in members of the same family. [8] However, the sister of the patient had normal kidneys, uterus and ovaries.

Renal agenesis has also been reported to be due to the teratogenic effect of diabetes mellitus, use of renin angiotensin inhibitors, high doses of vitamin A derivative, chlorambucil, and cocaine abuse. The exact etiology of the condition is still unknown. [2] The drug treatment for pregnancy induced hypertension and gestational diabetes mellitus can also be teratogenic with the consequence of renal aplasia or agenesis. [1] Yalavarty et al [10] stated that a pregnant woman with pre-conception diabetes mellitus has a 5-fold increased risk of having a fetus with unilateral renal agenesis. [10] The patient's mother had gestational diabetes mellitus which could have led to the introduction of teratogenic agents via the maternal disease or the drug treatment of the disease, to possibly produce a right renal aplasia or agenesis in the patient.

Assessment for the renal location prenatally is done in the 2nd and 3rd trimester. Post-natal assessment can be done at any time after birth. The sonographic appearance of the adrenal gland when the kidney is absent at its bed is the "lying down "adrenal sign. [13] Flattening of the adrenal gland and absent kidney at the renal bed is suggestive of unilateral renal absence or renal ectopia. Severe oligohydramnios in utero, is an important feature in unilateral renal agenesis. [2,4] The absence of a single renal artery using colour Doppler is also suggestive of the condition. In true congenital renal agenesis, the ipsilateral urinary bladder hemi-trigone will be absent. [10] The patient's right kidney was absent in the renal bed and the pelvic region, using ultrasound, but the urinary bladder was normal with no

history or complaints of abdominal distension due to urine leakage.

Plain film demonstrates a more medial position of the splenic flexure when there is left renal agenesis or of the hepatic flexure when there is right renal agenesis, whereas the bowel position is normal when the unilateral renal absence is due to an involution, atrophy and ultimate disappearance of a multicystic dysplastic kidney. [4] In the index case the hepatic flexure was normally sited, as seen in Fig.3.

In a bid to confirm if the kidney is ectopic, an intravenous urography can be done which will locate the presence of an ectopic functional kidney. In the present case there was no ectopic kidney detected. [1] Renal scintigraphy using Dimercaptosuccinic acid (DMSA) will also demonstrate the location of a functioning kidney if ectopic. [4,8] Magnetic resonance imaging can be utilized to demonstrate an ectopic non-functioning kidney and associated genital anomalies. [4]

Affected individuals should not be involved in contact sports like football, wrestling and martial arts. [4] Offspring of individuals with unilateral renal agenesis are at risk of more serious renal anomalies, therefore, ultrasound study of parents and siblings is recommended in all families with an affected individual. [7]

The affected individual is expected to be assessed yearly for the presence or absence of protein in urine, to check the blood pressure since they are prone to developing hypertension, and serum creatinine level evaluation. They are advised to minimize salt intake, increase water intake, maintain a healthy body mass index (BMI), take only drugs prescribed by a physician and restrict dietary protein intake if the creatinine clearance is less than 50%. [1,4]

CONCLUSION

An 18-year-old female who had right renal absence likely due to agenesis is presented. Ultrasonography demonstrated the absence of the right kidney in the renal bed and intravenous urography did not reveal the presence of a right ectopic kidney. She had been requested to visit the hospital for yearly assessment but she was still complaining of non-specific mild abdominal pains.

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