Unilateral Renal Agenesis Coexisting with Bilateral Cryptorchidism in an Adult Nigerian: Case Report

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SUMMARY

A case of unilateral renal agenesis discovered incidentally during an abdominal ultrasound scan to locate undescended testes in a 38 year old man is presented. There are very few reports about cryptorchidism in unilateral renal agenesis. In the general population cryptorchidism is seen in 4% of boys at birth decreasing to 1.8% at 1month of age and 0.8% at 9 months of age. This case is reported to highlight the association of the two conditions and to emphasize the importance of early diagnosis in order to institute treatment early to salvage the testis and preserve the solitary kidney from damage.

Niger Med J. Vol. 50, No. 3, July – Sept., 2009: 71 – 73.

Key words: renal agenesis, cryptorchidism, ultrasound, ureteric bud.

INTRODUCTION

Renal agenesis is a congenital anomaly of the kidneys in which there is absence of fetal kidney on one or both sides. Bilateral renal agenesis is incompatible with life and they are either still born or die soon after birth. Unilateral renal agenesis is compatible with life. The contralateral kidney is usually larger due to compensatory hypertrophy. The incidence of unilateral renal agenesis is 1 in 1000 autopsies.[1] Male to female ratio is about 1.8: 1.

It may be an isolated congenital malformation or may be associated with chromosomal abnormalities or a variety of non-chromosomal syndromes including VACTERL (V = Vertebral/ Vascular, A = Anorectal atresia, C = Cardiovascular, T = Traecho, E = Esophageal Fistula ± atresia, RL = Radial/ Renal) and MURCS (M = Musculoskeletal, UR = Urogenital, C = Cardiovascular, S = Skeletal) associations.[2] There have been reports of ipsilateral genital abnormalities in 20% to 70% of cases of renal agenesis.[3] Genital abnormalities associated with renal agenesis develop more frequently in males than females.[3] In the female, the most frequent abnormalities reported are biconuate or unicornuate uterus and vaginal duplication and atresia.[4] In the males, the main abnormalities include: hemitrigone assymetry, agenesis and cyst of the seminal vesicle, Wolffian paravesical cyst, cystic dilatation of the prostatic utricle and ectopic drainage of the ureter into the seminal vesicle. There are very few reports about cryptorchidism in unilateral renal agenesia. In the general population cryptorchidism is seen in 4% of boys at birth decreasing to 1.8% at 1month of age and 0.8% at 9 months of age.[5] Little has been published on the incidence and prevalence of cryptorchidism in Nigeria. Nwako recorded an incidence of 0.1-0.2% in young adults[6]; Okeke in a study at Enugu among primary schoolboys recorded a prevalence rate of eight per 1000.[5]

This case is reported to highlight the association of the two conditions and to emphasize the importance of early diagnosis in order to institute treatment early to salvage the testis and preserve the solitary kidney from damage.

CASE REPORT.

This is a case of 38 year old male oil company engineer who presented to his company clinic in town with a complaint of small scrotum. He was seen by a physician who subsequently referred him to the urology clinic of University of Port Harcourt Teaching Hospital. Patient is not married and the third of four siblings, two girls and two boys. There is no history of congenital anomaly amongst his siblings. There is no history of hypertension, diabetes mellitus or significant past history of ill health. Patient does not smoke but drinks alcohol occasionally. Physical examination revealed a young man, healthy looking, not pale, not jaundiced or cyanosed. There was no pedal oedema. Pulse rate was 80 beats per minute, regular and of good volume. Blood pressure was 120/80mmHG. Heart sounds 1 & 11 were heard. There was no murmur. Apex beat was at the 5th left intercostals space, midclavicular line. Respiratory, abdominal and central nervous system examination showed no abnormalities. The penile shaft was normal and the scrotal sac was devoid of testes. A diagnosis of undescended testes was made.

Abdominopelvic ultrasound (fig.1) scan revealed a solitary right kidney with empty left renal fossa. Search for the ectopic testis was unfruitful. Computed tomography scan axial (fig.2) and coronal reconstruction (fig.3) revealed a solitary large functioning, right kidney in normal position. Left renal fossa was empty. Both testes were not found. A diagnosis of unilateral left renal agenesis with bilateral cryptorchidism was made.

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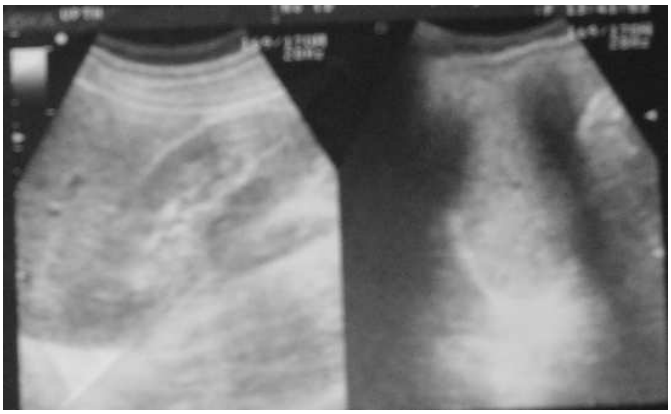

Figure 1: ultrasound scan images showing the right kidney in image A) and absence of kidney on the left in image (B). The black arrow points to the spleen.

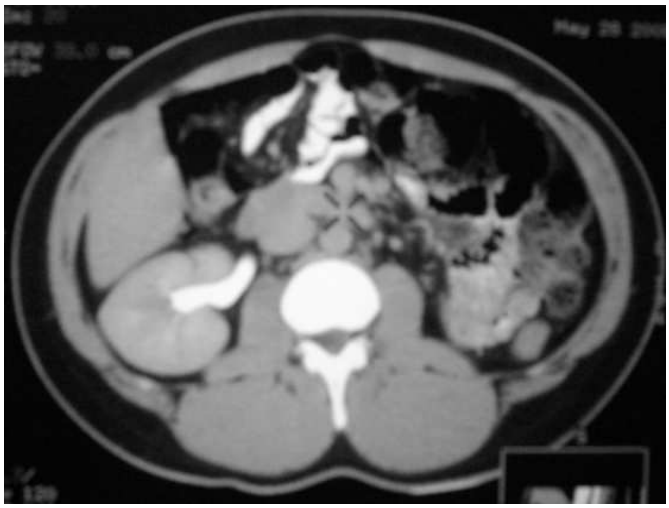

Figure 2: Axial CT scan of the abdomen at the level of the kidney showing absence of the left kidney. There is contrast media excretion in the right renal pelvis.

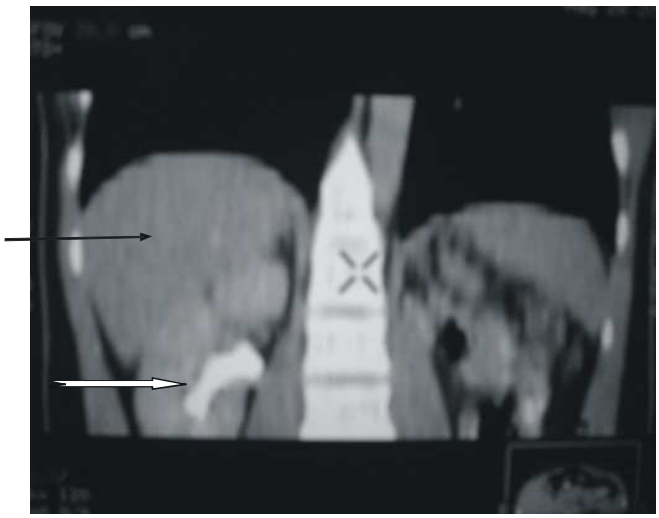

Figure 3: Coronal reconstructed CT image showing the right kidney (white arrow) below the liver (black arrow).

DISCUSSION

The urogenital systems originate in the intermediary mesoderm ocated along the posterior wall of the abdominal cavity between the fourth and the tenth week post-conception.[8] Alterations in this development before the fourth week can lead to unilateral agenesis of the genitourinary structures.[8] Renal agenesis is generally thought to result from lack of induction of the metanephric blastema by the ureteric bud which may be secondary to ureteric bud maldevelopment and/ or to a problem with the formation of the mesonephric duct leading to blind ending ureter.[9] It may also result from in-utero regression of multicystic renal dysplasia and from failure of the bud to form hemitrigone.[9]

Unilateral renal agenesis is usually asymptomatic and may be found incidentally during examination for congenital malformations, enuresis or abdominal pain, or patient could present with clinical features of renal failure.[10] Renal agenesis was discovered incidentally in this case reported in the process of investigating cryptorchidism. Nearly half (48%) of the patients with unilateral renal agenesis have associated urological anomalies which include vesicoureteral reflux (28%), ureterovesical junction obstruction (11%), ureteropelvic junction obstruction (7%) and coexisting ureterovesical and ureteropelvic junction obstructions (2%).[11] Vesicoureteral reflux is the most common associated anomaly with unilateral renal agenesis. None of these anomalies was found in this case presented. As a result of these possible coexisting anomalies it is advised that all patients with a solitary kidney should undergo a screening micturating cystourethrography and hysterosalpingography in females, as early recognition and treatment of these coexisting anomalies is imperative to decrease the long term risk of renal damage and infertility in affected patients.[10] There is increased risk of infertility, malignancy, testicular torsion and psychological impact in patients with cryptorchidism.[11] Unilateral renal agenesis is usually detected incidentally with ultrasonography and is confirmed using other radiological investigations as observed in our patient.

Intravenous urography or radionuclide scan using diethylenetriaminepentaacetic acid or dimercaptosuccinic acid may show the single kidney with its pelvicalyceal system. Abnormality in its size, position, alignment and function could be detected.[12] The trigone is usually deformed with the ureteral orifice missing on the involved side, so that cystoscopy may confirm the diagnosis. At times however, a portion of the lower ureter may be present in renal agenesis therefore the trigone may have no deformity.[10] In all cases of apparent agenesis, the lower abdomen and pelvis should be carefully scrutinized in order that a small ectopic kidney is not overlooked. On CT scanning, if the left kidney is absent, the renal fossa is filled by splenic flexure and pancreatic tail. On the right side, the fossa may be filled by duodenum, proximal small guts, hepatic flexure or liver.[12] CT may also detect presence of abdominal or pelvic testes. Venography is said to be more reliable than arteriography in making the diagnosis of renal agenesis.[13]

The diagnostic value of MR urography appears to be superior to that of other imaging modalities and is currently considered to be the gold standard for evaluating

sonographically suspected solitary kidneys[14] and associated vesicoureteral junction obstruction.[15]

In this case presented, ultrasound and CT scan were the radiological modalities used in the evaluation of the patient. MR urography and scintigraphic scanning were not used because they were not available at the time patient was being evaluated. Intravenous urography was not done because the primary concern of the referring surgeon was to localize the ectopic testes and the procedure was also considered unnecessary following CT image evaluation. Patient with unilateral renal agenesis and a normal solitary kidney are at increased risk of proteinuria, hypertension and renal insufficiency. Therefore it is essential to have prolonged and careful follow up and to employ strategies at maximizing renal preservation.[10] Since sonography is a non-invasive modality, it is an ideal tool for long term follow up for assessing the status of the solitary kidney.

CONCLUSION

A case of unilateral renal agenesis discovered incidentally during an abdominal ultrasound scan to locate undescended testes in a 38 year old man is presented. Early investigation of cryptorchidism or renal agenesis is very important in preventing the looming complications like renal failure and malignant transformation of the undescended testes.

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