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Crossed fused renal ectopia presenting as recurrent lower abdominal pain and urinary tract infection

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Abstract

Background: Crossed fused renal ectopia is a relatively rare condition which may remain undiagnosed for a long time. Renal function is usually preserved. It usually involves the left kidney. It is usually associated with mal-rotation, and may be a cause of urinary tract infection, hydronephrosis and renal calculi.

Material and method: We report a case of a 15-year old boy who was referred to our clinic with complaints of abdominal pain, and an ultrasound report of "absent right kidney".

Results: Urine examination indicated a partially treated infection, but the tests of kidney functions were normal. Intravenous urography confirmed crossed fused renal ectopia, and a flush aortogram showed two anomalous right renal arteries arising from the left side of aorta.

Conclusion: An "absent" kidney in its normal location should prompt further investigations. Recurrent chronic lower abdominal pain and urinary tract infection in a young person may be due to congenital renal abnormality. Evaluation of persistent urinary abnormalities in a child should include screening for congenital abnormalities.

Keywords: *Crossed fused renal ectopia. abdominal pain. recurrent urinary tract infection*

Résumé

Contexte: L'ectopie rénale croisée et fusionnée (la mauvaise fonction rénale) est une maladie relativement rare qui peut rester non diagnostiquée pendant une longue période. La fonction rénale est généralement préservée. Elle implique généralement le rein gauche. Elle est souvent associée à une mauvaise rotation, et peut être une cause d'infection des voies urinaires, hydronéphrose et des grains formulés dans le système rénal.

Méthode: Nous avons fait usage du cas d'un garçon de 15 ans qui nous a été transféré pour plaintes de douleurs abdominales, suivi du rapport d'échographie faisant cas "d'absence de rein droit".

Résultats: L'examen d'urine a indiqué une infection partiellement traitées, mais les tests de la fonction rénale étaient normaux. L'urographie intraveineuse a confirmé l'ectopie rénale fusionnée et croisée, et une aortographie a montré deux artères rénales droites anormales découlant de la partie gauche de l'aorte.

Conclusion:

Un rein "absent" à son emplacement normal devrait inciter aux investigations.

Les récurrentes douleurs abdominales chroniques et les infections des voies urinaires chez les jeunes peuvent être dues à une anomalie congénitale rénale. L'évaluation des anomalies urinaires persistantes chez un enfant devrait inclure le dépistage des anomalies congénitales

Introduction

Congenital anomalies of the kidney and urinary tract (CAKUT)are relatively common, occurring in 1 in 500 births [1]. Types of anomalies include agenesis, duplication, fusion, dysplasia, ureterocoele, and ectopia. Embryologically, ectopia of the ureter is the cause of many anomalies as diverse as renal hypoplasia, ectopia of the ureteral orifice, urinary outflow obstruction and reflux. The genetic causes (single-gene mutations and modifier genes) are complex, and in most case sporadic even though some may be familiar in nature. Genetic disorders of the rennin-angiotensin is also now thought to be involved in CAKUT [2]. Specifically, crossed fused renal ectopia occurs during the 4th to 8th week of gestation from abnormalities of the ureteric bud and metanephric blastema [3].The anomalies are mostly confined to the urinary tract [1].These anomalies cause renal failure and hypertension later in life if uncorrected. Therefore, recognition and early treatment of these conditions will reduce the substantial morbidity and mortality that they may lead to.

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Case report

A 15 yr old male student was seen on referral from another hospital as a case of 'absent right kidney with a suspicion of pelvic kidney and nephritis'. He had been complaining of recurrent lower abdominal pains for 2 years. The abdominal pains were colicky, usually exacerbated after exercise. There was no abdominal swelling and no constitutional symptoms. There was no previous abdominal trauma. He had microhaematuria and proteinuria.

When we saw the patient he had no complaints, especially of urinary symptoms such as dysuria and frequency, probably because he had been on antibiotics on several occasions. The lower abdominal pains were, however, recurrent. He weighed 52.1kg and his height was 1.74m, greater than 50th percentile for his age and sex for a Nigerian child [5], and also an evidence of his good nutritional status. He was not pale, anicteric and had no peripheral oedema. His external genitalia were normal. Cardiovascular, respiratory, and neurological examination did not reveal any abnormality. His blood pressure was 110/70mmHg. There was no abdominal mass or bruit.

A repeat abdominal ultrasound was done and this also reported 'absent right kidney, but the left kidney was normal' measuring 97.6mm by 37.9mm. No other abdominal organ abnormality was reported. Urinalysis revealed 3+ protein and 2+ blood. Urine microscopy showed numerous white blood cells but culture was negative. Urinalysis was negative for nitrite. Full blood count revealed a white cell count of 5.1 x 10^9/l, haemoglobin concentration of 13.3g/dl, neutrophils 52%, lymphocytes 43%, monocytes 3% and eosinophils 2%. Serum electrolytes, urea and creatinine are as follow: sodium 135mmol/l, potassium 4.2mmol/l, chloride 97mmol/l, bicarbonate 24mmol/l, calcium 1.99mmol/l, uric acid 0.387mmol/l and creatinine 58mmol/l. His 24-hour urinary protein was 0.09g.

Intravenous urography (IVU) was done and this revealed that there was no nephrogram on the right. There were double pelvis and ureters on the left, with the lower ends of the ureters overlapping the vertebra bone. There was no hydronephrosis and no evidence of calculi. The bladder was normal. The contrast was promptly excreted. A diagnosis of crossed fused renal ectopia was made. (Fig.1)

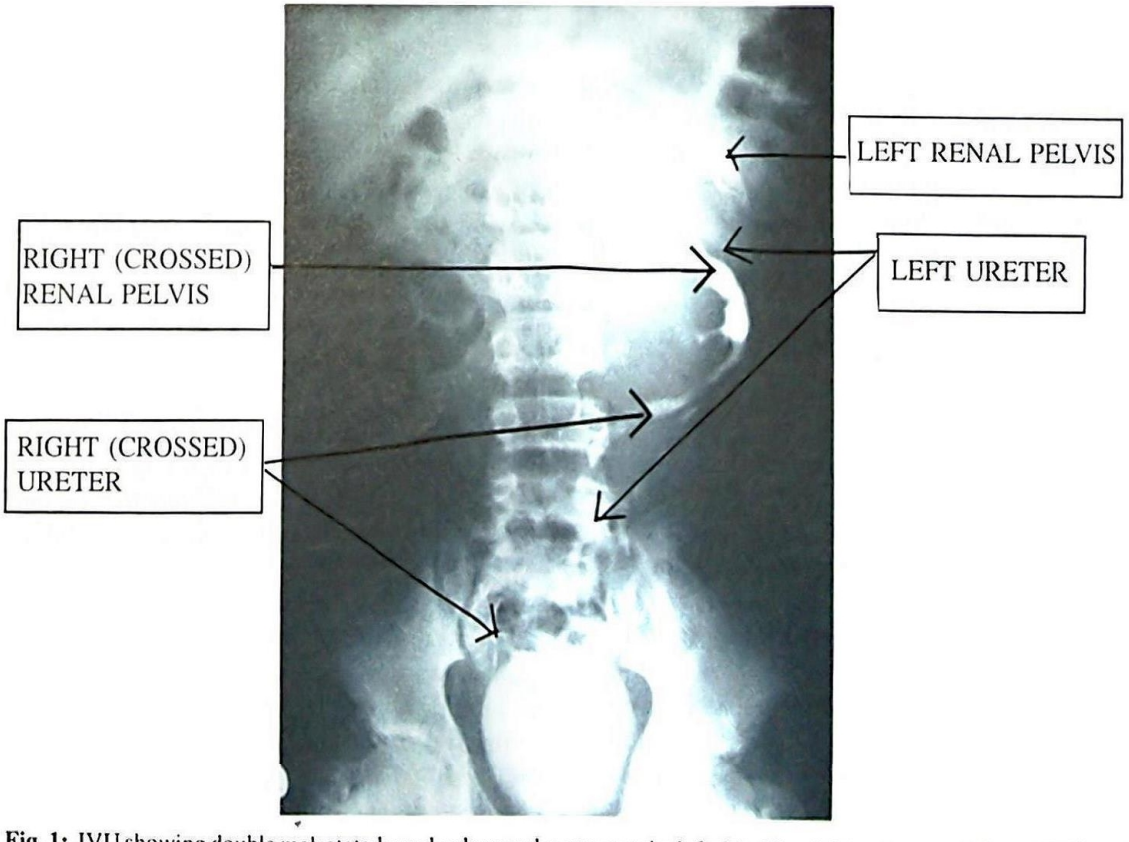

Fig. 1: IVU showing double malrotated renal pelves and ureters on the left side. The right ureter crossed over the lower lumbar and sacral vertebrae to insert into the bladder

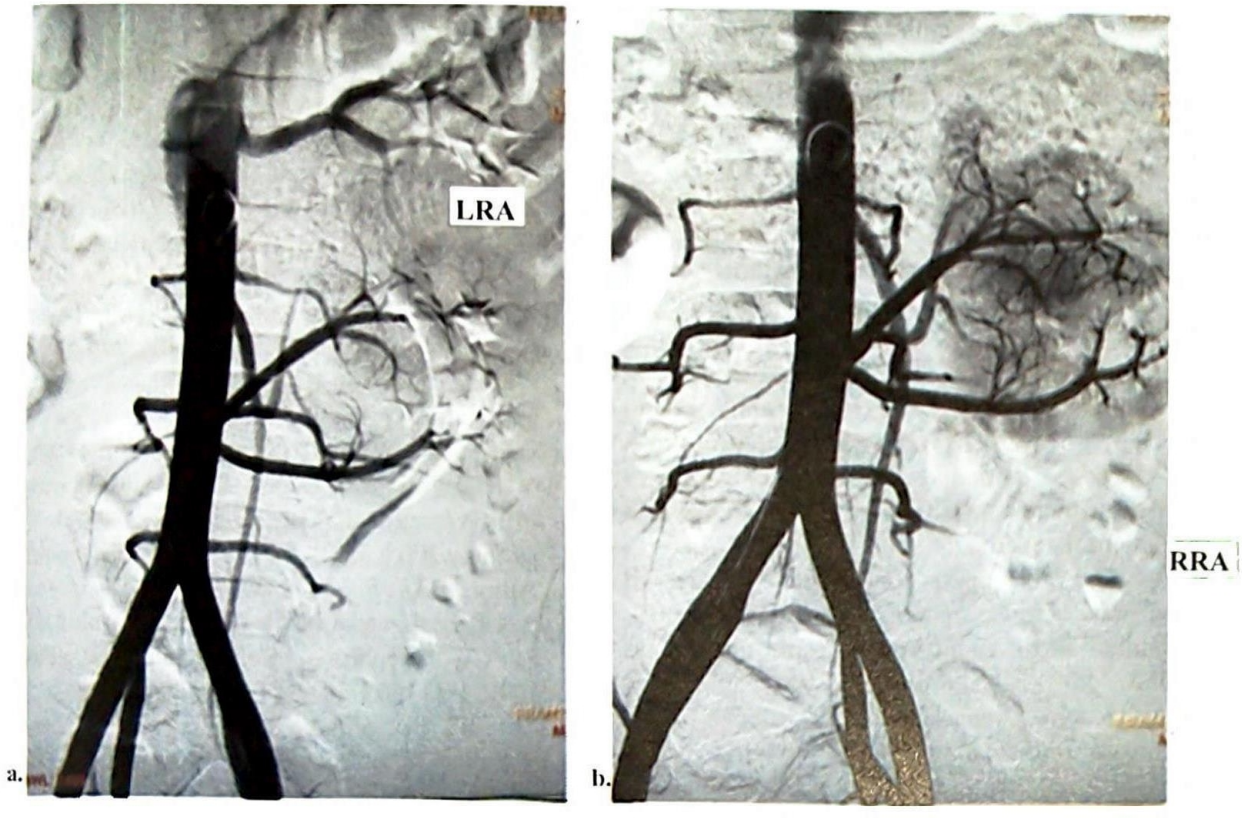

Fig. 2: (a). Flush aortogram showing normal left renal artery (LRA) from its origin at L1 vertebra. **(b).** There are two anomalous right renal arteries (RRA) arising from the left side of the aorta with a common origin at L3 vertebra.

A flush aortogram was done to exclude vascular abnormalities in case these were present and also to make early appropriate referral for follow up in this regard. This showed a normal left renal artery arising from its origin at L1. There were two anomalous right renal arteries arising from the left side of aorta with a common origin at L3. The arteries were normal in calibre and outline. They supply the crossed right kidney. They measured 37.8mm and 37.3mm in their respective lengths. The demonstrated nephrogram extended from the left hypochondrium to the lower lumbar region, 2.75 cm above the ipsilateral iliac crest. The excretion of contrast was prompt and good in both kidneys. The calyces were malrotated. The left ureter was longer, more lateral and was inserted into the left superior aspect of the urinary bladder; while the right crossed ureter was shorter, more medial and crossed the midline to insert into the right superior aspect of urinary bladder **(Figs.2a & b)** [^{99m}Tc] Dimercaptosuccinic acid (DMSA) and Diethylenetriamine penta-acetic acid (DTPA) are investigative techniques that could provide functional and structural information. Both were not done at this stage because his renal functions were judged to be preserved, and it was thought unnecessary to expose him to further radiation after the flush aortogram had been done. These may be done later if it is necessary to determine the contribution of each kidney to the overall renal function.

Based on the urine microscopy findings which suggested partially treated UTI, patient was treated with Co-amoxiclav. A repeat urine microscopy and culture was not remarkable after five days of antibiotic therapy. Patient has remained free of symptoms for three months and has been scheduled for a three monthly review in the clinic.

Discussion

Crossed fused renal ectopia is a relatively rare condition which may remain undiagnosed for a long time. Autopsy series estimate this to be 1 in 1000-2000. Renal function is usually preserved.

Ultrasonographic diagnosis may be difficult, and a crossed fused renal ectopia may appear as a renal mass [6]. It usually involves the left kidney [3], but in this case it is the right kidney that was involved. It is commonly associated with mal-rotation. The

crossed kidney usually lies below the normal kidney because it crosses the midline where its upper pole is fused to the lower pole of the normal kidney. Therefore, it may become complicated with hydronephrosis because of abnormal position and rotation of the ureters which may impede urine flow [7].For this reason, patient may develop chronic pyelonephritis from recurrent infection [8], calculi from stasis and reflux [9,10]. Usually, the crossed kidney does not ascend and rotate completely, and is then fused inferiorly to the normally located kidney [4]. This patient was treated for urinary tract infection (UTI), and, because of the colicky abdominal pain, we were also interested in excluding renal calculi especially when he had protein and blood in the urine. There was no evidence of renal calculi and we persisted with treating the patient for UTI from which the patient recovered.

The other major complications of crossed fused renal ectopia are renal carcinoma [11, 12], abdominal aortic aneurysm [13], and multicystic dysplasia [14]. Renal vasculature anomalies are a constant feature of renal ectopia [15] as demonstrated in this case. Demonstration of this by angiography is particularly necessary when surgery is contemplated in these patients.

Conclusion

An "absent" kidney in its normal location should prompt further investigations. Evaluation of recurrent lower abdominal pain and persistent urinary abnormalities in a child should include screening for congenital abnormalities.

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