CASE REPORT

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Horseshoe kidney presenting as recurrent urinary tract infections with severe hydronephrosis in a 10-month-old baby: a case report

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ABSTRACT

Introduction: Horseshoe kidney (HSK) is type of congenital anomalies of the kidneys and urinary tracts (CAKUT), which fusion anomalies happened in early embryonic period partially. One-third of the patients with horseshoe kidney are usually asymptomatic and diagnosed incidentally. It can be presented with other renal and extrarenal abnormalities. Here we report a case of horseshoe kidney in infant presenting as recurrent urinary tract infections with severe hydronephrosis.

Case Presentation: A 10-months old boy with multiple congenital anomalies admitted with a history of recurrent urinary tract infection for 3-month and progressive abdominal enlargement. His physical examination revealed an abdominal distention, no-tender, dullness on percussion, a palpable mass on the right flank, and normal bowel sound. The genitalia examination revealed penile-type hypospadia and bilateral undescended testis. Further examination with a contrast computed tomography scan showed HSK with severe right hydronephrosis with thin parenchyma due to UPJO. The heminephrectomy procedure was performed successfully. There is no recurrent UTI and no deterioration in renal function after 3 years of evaluation.

Conclusion: It is important to investigate renal abnormalities in patients with recurrent UTI. HSK is one of the prevalent findings. Therefore, proper diagnostic investigation tailored to the clinical presentation in a stepwise approach aiming for prompt and optimal patient care is essential.

Keywords: horseshoe kidney, pediatrics, severe hydronephrosis, urinary tract infection.
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INTRODUCTION

Congenital anomalies of the kidneys and urinary tracts (CAKUT) are disorders caused by defects in the development of the kidneys and their outflow tracts. The prevalence of CAKUT is estimated at 4–60 per 10,000 births. Congenital kidney malformations are defined macroscopically by changes in kidney size, shape, position, or number, or microscopically by a reduced number of nephrons and/or abnormal histology.[1,2] Horseshoe kidney (HSK) is type of CAKUT, which fusion anomalies happened in early embryonic period partially.[3] It is occurring in 1:400-800 live births and more common in males.[4]

The development of the kidney begins in the 4th week of gestation by inductive interaction between the ureteric bud and the metanephric blastema. Congenital renal anomalies can occur due to abnormalities of development, migration and rotation. The precise mechanism of the development of renal fusion anomalies is not fully understood and several theories have been put forward to explain the anomaly. The Mechanical Theory proposes that during cephalad migration, the kidneys pass through the fork between the two umbilical arteries and any positional change in these arteries squeeze the kidneys close together allowing their fusion (resulting in HSK).[3]

One-third of the patients with HSk are usually asymptomatic and diagnosed incidentally. It can be presented with other renal (46,1%) and extrarenal abnormalities (50%).[5,6,7] HSK cause the drainage to malfunction in the collecting duct system and may cause the formation of urinary stasis and renal stones.[8,9] This situation increases urinary tract infection (UTI) risk in patients with HSK anomaly. It was reported that urinary stasis and renal stone-related UTI prevalence in patients with horseshoe kidneys anomaly was 27% and 42%. Genitourinary or systemic additional anomalies may frequently accompany the clinical picture in the patients with horseshoe kidneys anomaly. The most prevalent accompanying urinary system anomalies are renal stones, vesicoureteral reflux (VUR), ureteropelvic junction obstruction (UPJO) and hydronephrosis.[8,10,11] Here we report a case of horseshoe kidney in infant

with severe hydronephrosis presenting with recurrent urinary tract infections.

CASE REPORT

A 10-months old boy was admitted to Prof DR I.G.N.G. Ngoerah Hospital with a history of recurrent UTI (four times) in 3 months preceding progressive abdominal enlargement. He had a fever and chill and had already gotten several oral antibiotic therapies (amoxicillin and cefixime). He also had progressively abdominal enlargement without complaints of pain and vomiting. The patient's parents said there were no disturbances or changes in eating, drinking, urinating, or defecating patterns. There are no members of the family with similar complaints. He had a medical history of multiple congenital anomaly (hydrocephalus, polydactyly) and congenital hypotiroid.

Upon evaluation, the patient is alert with normal vital sign. His physical examination revealed an abdominal distention, no-tender, dullness on percussion, a palpable mass on the right flank, and normal bowel sound (Figure 1). Genitalia examination found penile-type hypospadia and bilateral undescended testis.

Laboratory findings revealed normal blood count, liver enzymes, blood glucose, electrolyte serum, and creatinine serum (0.35 mg/dL) with an estimated glomerular filtration rate (eGFR) was 135 ml/min/1.73 m^2. However, his urinalysis showed normal with a negative urine culture.

Ultrasound evaluation of the abdomen revealed right hydronephrosis with unvisualized ureteral structures suspicious of a UPJO. Kidney enlargement passes through the midline into the pelvic cavity with the thinned cortex and pelvicalyceal system looking severe ectasis. There are no stones or masses seen. The visualization of the left kidney is within normal limits. (Figure 2).

The abdominal contrast computed tomography (CT) scan showed HSK with severe right hydronephrosis with thin parenchyma due to UPJO (Figure 3) and a normal left kidney joined by an isthmus. A micturating cystourethrogram (MCUG) examination showed no abnormalities (Figure 4).

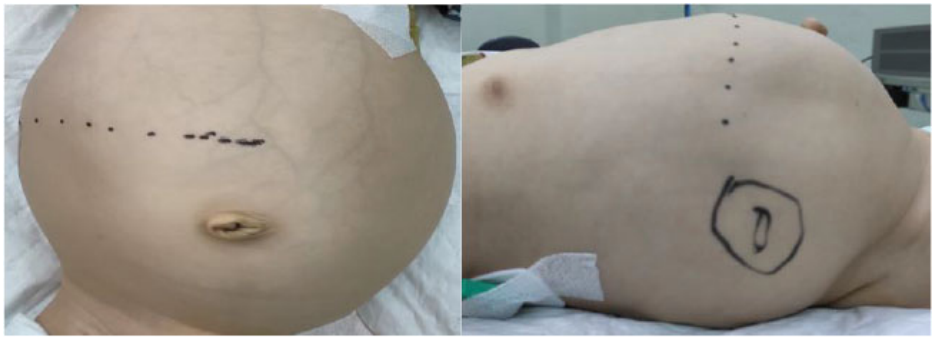

Figure 1. Patient's abdominal distention with 51 cm abdominal circumferential.

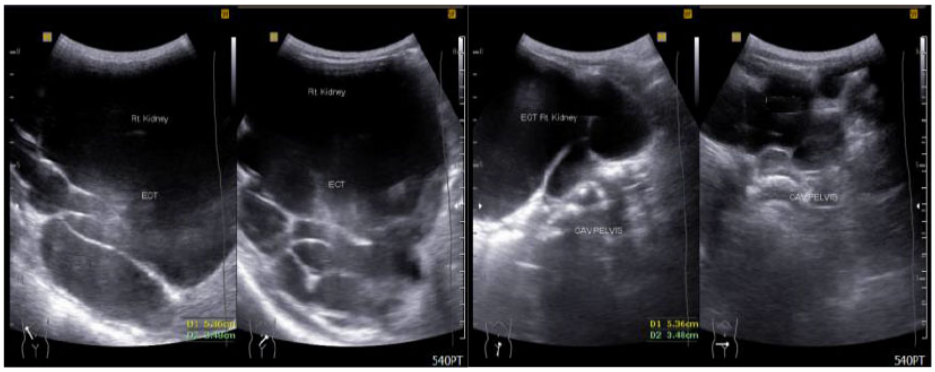

Figure 2. Ultrasound revealed severe hydroureteronephrosis on right kidney.

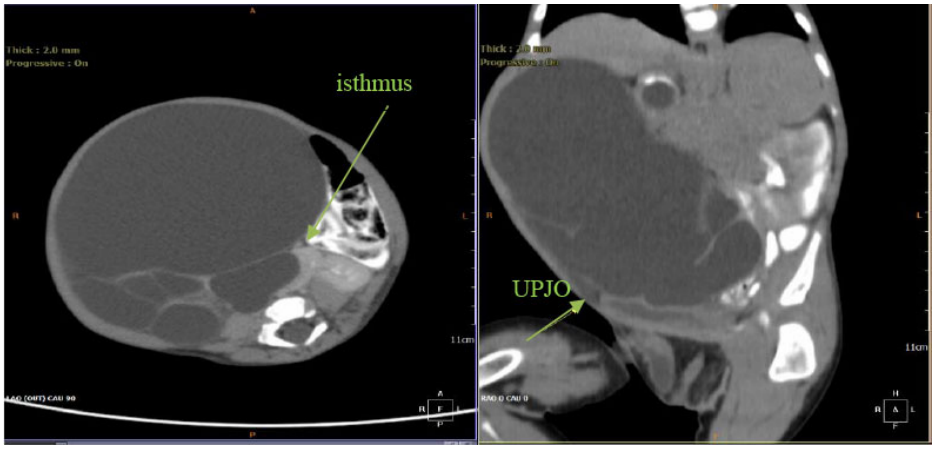

Figure 3. CT abdomen showing Right severe hydronephrosis with UPJO.

The operation was performed with a transverse upper abdominal transperitoneal incision. At the time of exploration, it revealed a dilatation of the right renal pelvis and obstruction 3 cm distal to the ureteropelvic junction (Figure 4). A right nephroureterectomy was performed successfully.

Postoperatively, urine production and a glomerular filtration rate (GFR) were within normal limits. The histopathological revealed chronic pyelonephritis. The patient made an uneventful post-operative recovery. There is no recurrent UTI or deterioration in renal function (creatinine serum 0.35 mg/dL with eGFR was 107.8 ml/min/1.73 m^2) after 3 years of evaluation (Figure 5).

DISCUSSION

Horseshoe kidney starts between weeks 4 and 6 of gestation when the inferior part of the metanephric blastema fuses before the kidneys ascend and rotate on their long axis. It forms an isthmus of connective or functional tissue. The isthmus prevents the kidneys from ascending and rotating

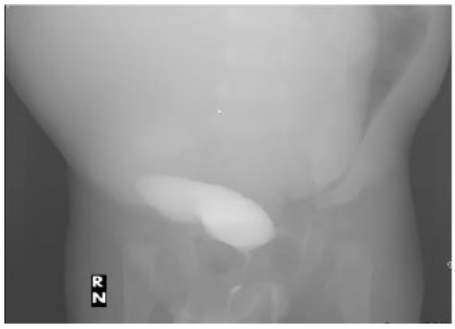

Figure 4. It was a negative result of vesicoureteral reflux on a cystogram.

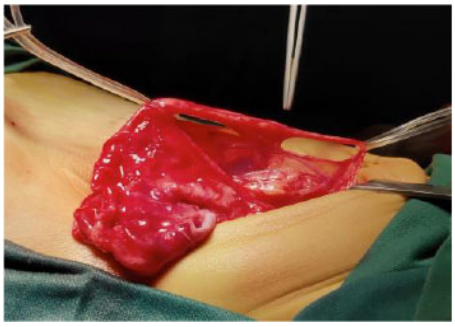

Figure 5. ureter obstruction was found during operation.

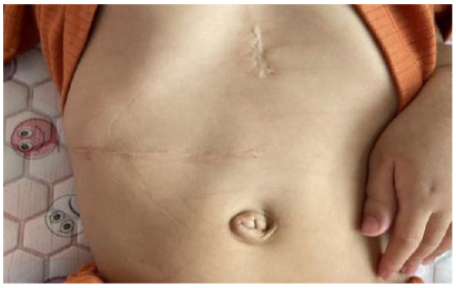

Figure 6. Patient's abdomen after 3 years of surgery.

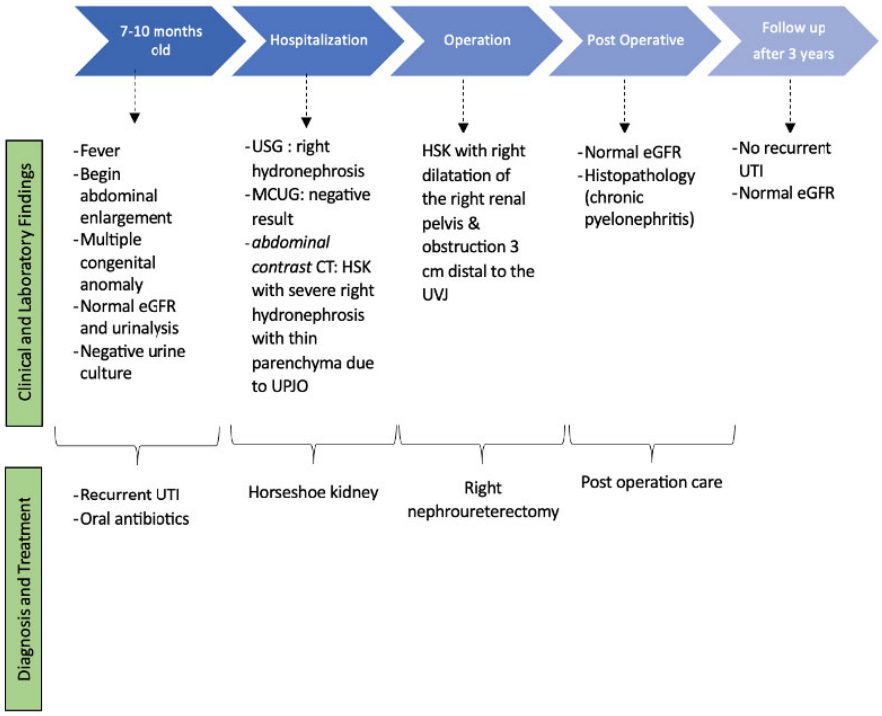

Figure 7. Case report timeline.

due to the resistance when it reaches the inferior mesenteric artery. HSK usually consists of renal ectopic, malrotation of the kidneys, and renal vascular abnormalities. In most cases of HSK, the kidneys connect by an isthmus located at their lower pole, and in fewer cases, the isthmus connects to the upper pole.[5] In many cases, the HSK can be found on ultrasound or intravenous urography (IVU). It is sometimes found on a plain radiograph as the renal axis is altered with the lower poles being more medial than expected. The kidney may be seen to be lower in position compared with the normal. However, plain film radiography is too insensitive to make a diagnosis and another imaging method is required. Ultrasound can help in direct visualization of the isthmus and also show the location and abnormal orientation of the horseshoe kidney. However, it is not sensitive in patients with large body or in cases of horseshoe kidney with fibrous isthmus. In addition, ultrasound is operator-dependent and the detection rate varies depending on the skill and experience of the operator. Contrast-enhanced CT is the modality of choice for evaluation of the horseshoe kidney and relation to surrounding structures. It also plays an important role in the evaluation of potential complications and surgical planning. This allows for the identification of stones as well as urinary blockages and UPJO. MRI can be used with similar advantages and without the risk of radiation, however, some complications including stones and trauma are better evaluated with CT.[8,12] In this case, the HSK came from an anomaly fusion of the inferior pole detected by a CT scan procedure.

Horseshoe kidney may be diagnosed at any age ranging from the fetus to the elderly patient. It is most commonly diagnosed in children because of its association with other congenital anomalies.[13] It may be present with several complications including renal stones, UPJO, hydronephrosis, VUR, urinary tract infection, malignancies and, loss of renal function. It also occurs with other congenital anomalies like cardiovascular, gastrointestinal, and central nervous system malformations, undescended testis and hypospadias, and chromosomal abnormalities.[5,8,10,11] In this case, HSk was discovered in a 10-months old male with a history of recurrent UTI and severe hydronephrosis, with multiple congenital anomalies including hydrocephalus, polydactyly, penile-type hypospadias, and bilateral undescended testis.

Urinary tract infection is one of the most common complications in HSK. Prevalence of UTI in patients with horseshoe kidneys anomaly in the literature was reported between 27%-42%; this rate is significantly higher in comparison to the UTI prevalence in healthy children.[9] Clinicians diagnose patients with UTI comprehensively based on clinical conditions, collect urine and blood samples, and begin broad-spectrum antibiotic treatment. Later, when the diagnosis of UTI is more definite based on the culture results, targeted antibiotics will be prescribed. However, the pathogenic

bacteria remains unidentified if the culture test is negative. This is potentially due to previous antibiotic exposure, error during sampling, an insufficient sample volume, or unculturable bacteria. Unidentified pathogenic bacteria can cause many issues in the quality of diagnosis and treatment, such as the administering less effective antibiotics, and futile repeated antibiotic administration.[14] In this case, the patient's urine culture was negative due to a history of consumption of oral antibiotics before. We still diagnose UTI based on clinical symptoms and predisposing factors (HSK).

Given that HSK is prone to various complications, the earlier it is detected the better. It is important to know if age is associated with the occurrence of complications. HSK is related to urologic and nonurologic abnormalities. Half of children with HSK have renal complications or extrarenal diseases or syndromes, with the incidence is higher in younger children.[15] Although HSK is often asymptomatic, sometimes it can lead to various complications. There is a lack of data on the long-term outcomes of children with HSK. According to the report, 80% of children with HSK develop hydronephrosis. VUR, UPJO, or an external compression of the collecting system by an aberrant vessel or renal calculi might cause hydronephrosis. UPJO is thought to be the result of the high insertion of the renal pelvis and displacement of the fused isthmus. HSK is particularly predisposed to infection that occurs as a result of VUR, stasis, or stone. UTI has been found in up one-third to one-half of the patients with HSK. In this journal, it said within the follow-up to 10 years, proteinuria and hypertension were detected in 15% and 10% of patients, respectively. Almost 7% of cases progressed to chronic kidney disease (CKD). The presence of proteinuria, hypertension, and renal scarring was found to be an independent risk factor for developing CKD in HSK.[16]

Management of patients with HSK is depends on symptoms and clinical presentations. In UPJO associated with renal anomalies, pyeloplasty or uretero-uretero anastomosis can be performed for the preservation of kidney function. Meanwhile, in cases where the kidneys have experienced severe dysplasia and are no longer functioning, nefroureterectomy is the best choice.[15,16,17] In our patient, right nefroureterectomy was performed because of severe right hydronephrosis with very thin cortex of the renal parenchyma. Renal function was preserved in the patients and all symptoms resolved post heminephrectomy.

CONCLUSION

It is important to investigate renal abnormalities in patients with recurrent UTI who have congenital abnormalities. HSK is one of the prevalent findings. Therefore, proper diagnostic investigation tailored to the clinical presentation in a stepwise approach aiming for prompt and optimal patient care is essential. We report the successful surgical management of a boy with recurrent UTI and progressive abdominal distension due to severe hydroureteronephrosis of the right side of HSK.

CONFLICT OF INTEREST

The authors declare that there is no competing interest regarding the manuscript.

ETHICAL CONSIDERATION

The patient's parents has signed a written informed consent and agreed to this study's publication.

FUNDING

None.

AUTHOR CONTRIBUTION

All authors equally contribute to the study by selecting case, evaluating the outcome, and reporting the study results through publication.

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