CASE REPORT

Pancake Kidney: A Rare Developmental Anomaly in Child

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ABSTRACT

The aim of the study is to present a rare case of incidentally detected pelvic pancake kidney with radiological imaging findings in a 11 year old male child .There are many developmental anomalies of kidney Various types of congenital renal abnormalities such as renal agenesis, ectopic or horseshoe kidney can be encountered where one of the most rarest presentation with a single ureter on pancake kidney is encountered in number that count in fingers worldwide.

Keywords: Pancake kidney, Renal Ectopia, Rare anamolies of kidney.

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INTRODUCTION

Wilmer in 1938 was the first to describe the logical categorization of fusion anomalies of the kidney while McDonald and McClellan in 1957 refined and expanded the classification given by Wilmer.[1] The classic features seen is Disc, shield, doughnut, or pancake kidneys are kidneys that have joined at the medial borders of each pole to produce a doughnut or ring-shaped mass. More extensive fusion along the entire medial aspect of the kidneys creates a disc or shield shape. Pancake kidney malformation results from complete medical fusion of the metanephric blastema at an early stage of embryonic development and is characterized by a single, flat, non rein form mass, in a medial position within the pelvic cavity or at the level of aortic bifurcation. The renal collecting system is anterior and typically drains with two ureters or less commonly via a single ureter. Vascular of such presentation is also anomalous i.e blood flow can be supplied by multiple branches of the internal and external iliac arteries or of the branches from abdominal aorta.[2]

CASE REPORT

A 11 year male child presented to urology outpatient department with history of on / off periumblical region pain for 15-20 days. Child complains of pain during school hour mild dull aching pain, non-radiating, no any aggravating factors, which relief during rest at home. Bladder habit is normal. He denied fever, burning micturation or loss of weight and appetite. He is active, well rehydrated, with unremarkable abdominal examination. Groin examination shows right scrotal swelling with cystic consistency suggestive of congenital hydrocele. Laboratory data were unremarkable patient is advised for abdominal and pelvis ultrasound to R/o mesenteric lymphadenitis and ultrasound imaging of the abdomen and pelvis demonstrated ectopic lobulated pelvic kidney located in the midline demonstrating hypoechoic cortex and hyperechoic medulla with adequate corticomedullary differentiation. Plain and contrast enhanced CT & CTU scan was performed showing empty B/L Renal fossa with ectopic lobulated pelvic kidney located in prevertebral region , slightly to the right of the midline showing normal opacification on corticomedullary phase and normal excretion on excretory phase. Single renal artery originating from the abdominal aorta is seen supplying the ectopic kidney with a single

renal vein and draining into IVC. Single renal pelvis (located in postero-medial aspect) along with a Single and slightly prominent short ureter is seen draining the kidney.

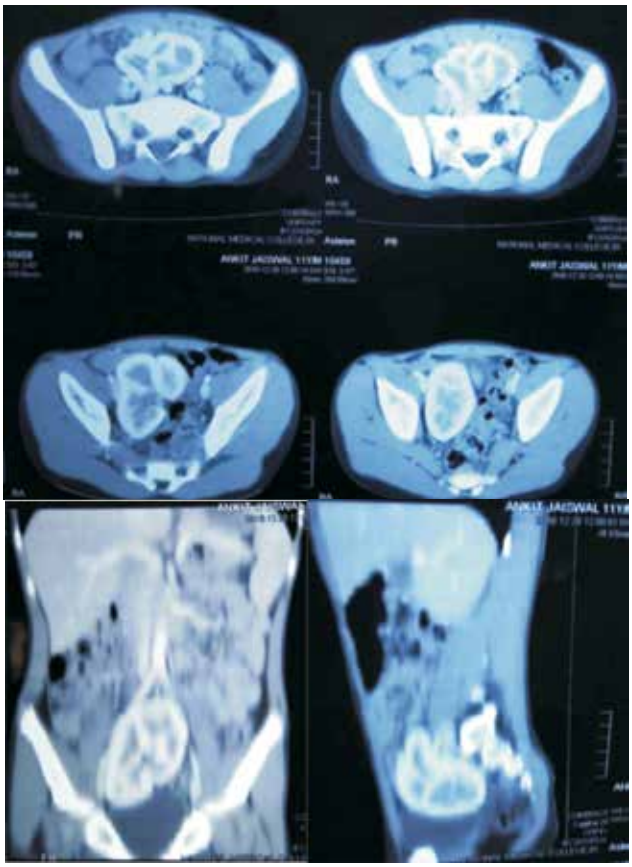

Figure : CT view of Pancake kidney (Custody: Radiology Department, National Medical College, Birgunj, Nepal)

DISCUSSION

Pancake kidney with cysts and a single ureter is rare type that was presented from Brazil showing asymptomatic cases however in our case patient present with pain abdomen bring us towards diagnosis of a new case with rare anamolies.[3] Cases associated with nephrolithiasis, hydronephrosis and vesicoureteral reflux resulting to recurrent urinary tract infection is in literature. Most cases with pancake kidney have anomalous rotating collecting system with short ureter leading to stasis and obstruction. However case report up to 65 years age is in literature so survival with single ureter and pancake is more than 5th decades of life. Case of 19 years female with lower abdomen pain for 2 months with fever and burning micturation, she has uncrossed ureters opening separately in to urinary bladder. However recent review with a prenatal ultrasonography detected incidence of 0.003% of crossed renal Ectopia in 1 case out of 400 cases of autopsy and 85% of them occur with a fused kidney , among them horseshoe kidney is commonest with incidence of 1 in every 700 autopsies.[4] Our report is of male child who is 11 years of age now with single ureter in comparison of 56 years old male with single ureter, survival is still high. Although much little case around 20 cases is reported among which single ureter is rare in fact with less than 10 cases have these findings. In the literature there have been cases of fused pelvic kidney reported to have concomitant anomalies such as Fallot tetralogy, vaginal absence, sacral agenesis and caudal regression.[5] Chromosomal Syndromes Associated with Genitourinary Anomalies the series of cases is seen with 9p trisomy, 9p tetrasomy, Strabismus, Pancake kidney and undescended testis however in our case patient present with right sided congenital hydrocele which was quite different. In literature it is also found that abnormally located umbilical artery may force the metanephric masses in to opposition and cause fusion and after the fusion occurs cranial ascent to the lumbar position is impaired by the retroperitoneal structures.[6] However supply of pancake kidney is from abdominal aorta and iliac artery above mentioned fusion because there is not well known incidence of the cases such integration to the cause of umbilical artery forcing fusion is debatable. Fact is that early identification of renal abnormalities is much important to evaluate associated risk of urinary system in future and its prevention. Prenatal USG and Post natal CT (KUB + IVU) are the investigations of choice for detection.[7]

CONCLUSION

In developing world like our setup ultrasound is not enough to detect all fundamentals of the pancake kidney in prenatal period. Testing renal function is not adequate for the excretory function needed to know whether single or double ureter are there and vascular status whether it is complicating compressing aortic bifurcation or not. But it shows survival of patient is confirmed up to 6th decades after looking incidence from child to geriatric age group however other risk factors and mortality are not well studied due to fewer amounts of cases and variation in diversity according to geographical regions including developed world.

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