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OBSTRUCTED PAN CAKE KIDNEY IN A CHILD MANAGED BY IMAGE GUIDED PERCUTANEOUS NEPHROSTOMY

Sheo Kumar[1], Prabhaker Mishra[2], Swish Kumar Singh[3]

[1]Professor, Department of Radiodiagnosis, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow-226014, India

[2]Assistant Professor, Department of Biostatistics & Health Informatics, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow-226014, India

[3]Senior Resident, Department of Radiodiagnosis, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow-226014, India

ABSTRACT

Pancake kidney, also called pelvic fused kidney, lump or cake kidney, is a rare type of congenital renal fusion anomaly. It is characterized by the presence of a lobulated pelvic renal mass which has a dual parenchymatous system without an intervening septum. Pancake kidney is prone to obstruction due to abnormal rotation of collecting system and calculus. When a patient presents with a nonfunctioning and obstructed kidney, a temporary nephrostomy preserves any recoverable renal tissue. Many a times a seemingly alarming radiological appearance where little functional recovery is expected, dramatic improvement in renal function has been noticed. We report a case of pancake kidney with right PUJ and left ureteric calculus causing hydronephrosis in a 9-years-old male child, evaluated by ultrasonography and MDCT, underwent image guided (sonography and fluoroscopy) percutaneous nephrostomy for relieving the ureteric obstruction.

Key words: Renal fusion, pancake kidney, percutaneous nephrostomy, nephrostogram.

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***Address for Correspondence:**

Prof. Sheo Kumar, Department of Radiodiagnosis, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow-226014, India

INTRODUCTION

In renal fusion anomalies which both kidneys are fused together in early embryonic life. They may be partial or complete. Partial renal fusion is represented by the horseshoe kidney and crossed renal ectopia with fusion. Pancake kidney is an anomaly characterized by the complete fusion of both kidneys. The term pancake kidney or fused pelvic kidney was defined by Glenn as an anomaly, in which "the entire renal substance is fused into one mass. Pancake kidney is a rare congenital anomaly of the genitourinary system, with fewer than 30 cases described in the literature [1]. Looney and Dodd were the first to describe pelvic pancake kidney [2]. The early diagnosis of potential complications like urinary tract infection, stone formation and obstructive uropathy that can accompany this anomaly must be made in order to prevent permanent renal damage [1, 2]. Percutaneous nephrostomy is a procedure of establishing a drainage tract into the upper urinary system by puncturing the kidney directly through the skin [3]. Percutaneous renal access can be achieved under fluoroscopic control or using an ultrasonography (US)- guided puncture [4]. We performed unilateral left sided percutaneous nephrostomy (PCN) which helped in confirming the diagnosis as evidenced by contrast opacification of pelvicalyceal system of both renal components. Unilateral PCN allowed drainage of bilateral obstructed system in one go. Ultrasound, NCCT KUB region and MDCT nephrostogam findings also discussed.

CASE REPORT

A 9 year old male child presented with fever, reduced urine output, dysuria, nausea and vomiting and edema all over the body for one week. In view of fever, leukocytosis and urine analysis s/o UTI, empirical antibiotics were started. S. creatinine and B. urea were

5.3mg/dl and 69.5 mg/dl respectively. One session of hemodialysis was given on the day of admission as there was absolute anuria with raised S. creatinine and volume overload. Urgent sonography was done which showed bilateral hydronephrosis and low position with some fusion of kidneys.

Contrast CT study could not be done as patient was having high S. creatinine levels. So NCCT was performed which showed fused kidneys as a single mass in midline in lower abdomen and upper pelvis. There was a calculus in right pelvi ureteric junction and another in left upper ureter with hydronephrosis in both renal components **(Figure 1: a, b).** Patient was immediately taken up for left percutaneous nephrostomy.

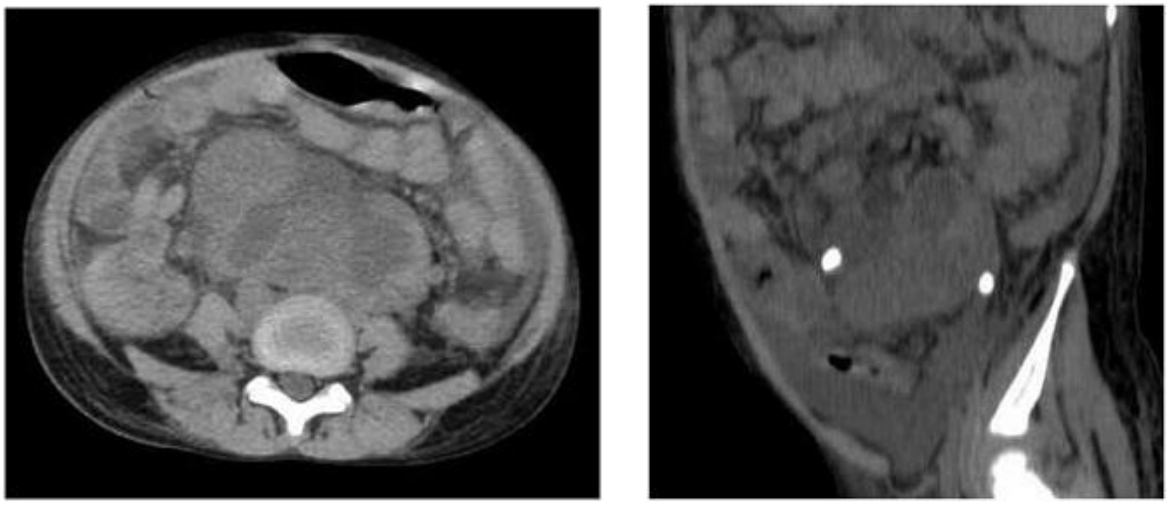

Figure 1 a & b: NCCT axial image (a) showing fused kidneys as a single mass and reformatted coronal oblique image (b) showing calculus in right pelvi-ureteric junction and in left upper ureter with hydronephrosis in both renal moiety.

Patient was put in prone position and a supporting pillow was placed under the abdomen. Under strict aseptic precaution and under local anesthesia, sonographic (Sonosite Micromax portable ultrasound machine with 2-5MHz probe) and fluoroscopic guidance (Philips C-arm impulse fluoroscopy machine) left renal moiety upper calyx was punctured with 22G chiba needle and metallic guide wire was passed.

Then 4F Neff percutaneous access canula was passed and guide wire was exchanged to 0.035", 150cm Terumo guide wire (Terumo Europe N.V, Belgium). Then the tract was dilated with 10F Coon's taper dilator (Cook Medical, Bloomington, USA) and finally 10 F percutaneous malecot nephrostomy catheter (Devon innovation Pvt Ltd, HP, India) was placed just proximal to right renal moiety pelvic calculus. Thick turbid white color urine (pus) was drained out.

Fluoroscopic nephrostogram and CT nephrostogram were performed after injecting contrast media through the percutaneous nephrostomy catheter, which showed contrast opacification of pelvicalyceal system of both renal moieties **(Figure 2 :a, b, c & Figure 3).**

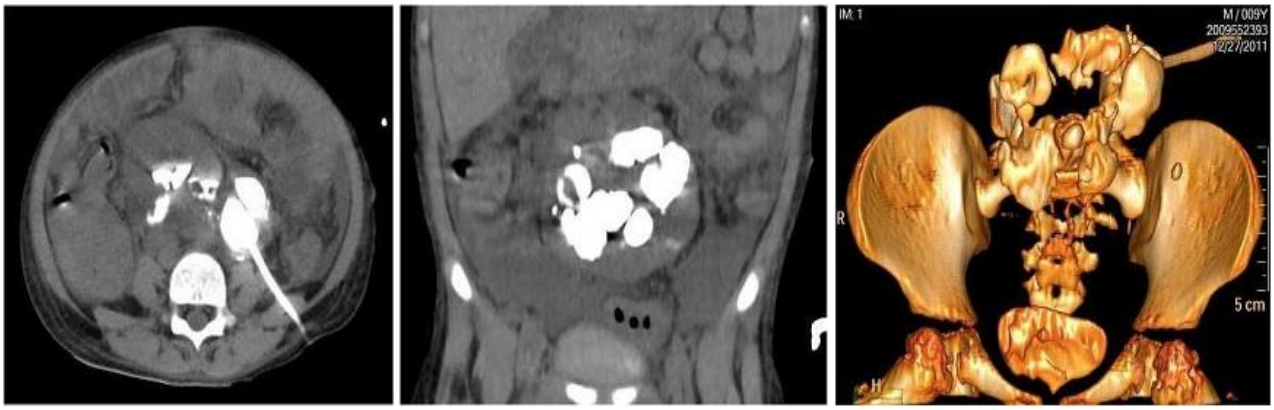

Figure 2 a , b and c: CT nephrostogram through left sided percutaneous nephrostom -axial (a), coronal (b) and volume rendered (c) images showing contrast opacification of pelvi-calyceal system of both renal moiety.

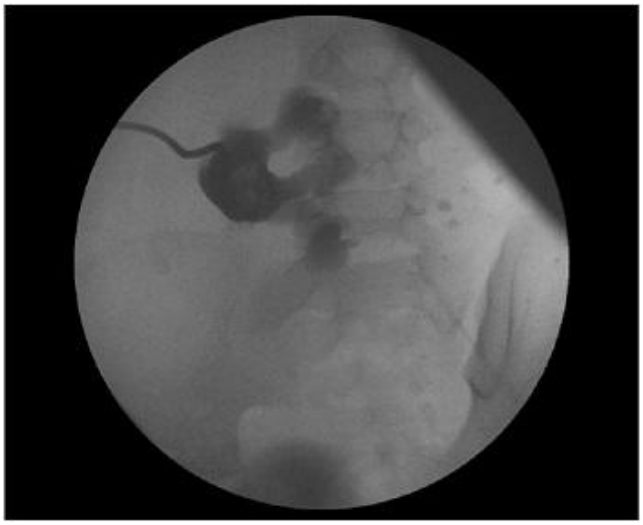

Figure 3: Fluoroscopic nephrostogram through left sided percutaneous nephrostomy showing contrast opacification of pelvi-calyceal system of both renal moety.

The catheter drained 200ml of pus followed by urine output of 4000ml in 12 hrs. Per-urethral urine output was less than 100ml. Urology consultation was sought. They planned for definitive treatment at a later date till present condition stabilizes. Serum creatinine and Blood urea levels were reduced. Patient was discharged in stable condition with advice to continue antibiotics. Unfortunately patient did not turn up for follow up. Although urinary system anomalies often coexist with malformations of other organs and systems, no associated anomalies were detected in our case.

DISCUSSION

In embryological life, two metanephric tissues occur in the pelvis which later shows cranial ascent, axial deflection, medial rotation and lateral migration. The embryological basis for pancake kidney is fusion of each metanephric mass in the pelvis during early ascent. An abnormally located umbilical artery may force the metanephric masses into opposition and cause fusion. After fusion occurs, cranial ascent to the lumbar position is impaired by the retroperitoneal structures. The vascular supply of the pancake kidney is consistent with its arrested ascent and derives from the common iliac artery or terminal aorta. Histologically, fused pelvic kidney shows cystic changes, immature glomeruli and dilated tubules.

Pancake kidney is more common in men [5]. The fused kidney occupies prevertebral or presacral space [1]. In the literature there have been cases of fused pelvic kidneys reported to have concomitant anomalies such as Fallot tetralogy, vaginal absence, sacral agenesis and caudal regression [2], failure of testicular descent, spina bifida, and anal abnormalities [1].

The presence of a pancake kidney may predispose to recurrent urinary tract infections and stones. This is due to the probable rotation anomaly of the collecting system and short ureters which are prone to stasis and obstruction [2]. The early diagnosis of complications that can accompany this anomaly must be made to prevent permanent renal damage [1]. Sonographic and CT findings were efficient, not only in detection and evaluation of pancake kidney anomaly, but also in exclusion of concomitant anomalies as well [2].

Obstructive uropathy is a potentially life threatening condition and sometimes it is desirable to provide immediate temporary relief of the obstruction, until definitive treatment can be undertaken. Percutaneous nephrostomy is a procedure of temporary drainage of an obstructed kidney by establishing a drainage tract into the pelvicalyceal system of the affected kidney directly through the skin [3]. PCN was first described by Goodwin et al. in 1955 [6] and recently reviewed by Dyer et al [3].

Classically it involves fluoroscopy guided puncture followed by tract dilatation and insertion of the tube with or without use of guide wire and a success rate of around 95% has been reported [3]. The shortcomings and side effects of extensive radiation during therapeutic procedures are well known. The choice of method for the type of access depends on training and personal preference. The advantages of US-guided puncture are avoidance of radiation, avoiding adjacent and visceral injury and, most importantly, intrarenal vascular injury [4]. The first ultrasonography-guided percutaneous nephrostomy was performed by Pederson and achieved a success rate of about 70%. Since then, a large number of studies of ultrasound guided percutaneous nephrostomies have been carried out, particularly in the last two decades and a success rate up to 92% have been reported, which is comparable to the fluoroscopy-guided methods without any appreciable radiation hazard. In a dilated system the posterior lower pole calyx is easily identifiable from a below 12th rib approach in a normally positioned kidneys. In an abnormally located kidney posterior lower pole calyx was identified sonographically. Higher calyces are more difficult to be discerned individually. Locating and puncturing a dilated calyx is straightforward but correct placement of catheter could be a bit difficult [3].

Although a pigtail catheter gives better patient tolerability, it is not ideal in situations where blood,

mucus, pus or stone is expected to pass because of its small caliber. Malecot tubes are preferred as they provide large-bore drainage [4].

Percutaneous nephrostomy can be performed on an outpatient basis in selected patients. Patients who live alone or in whom the risk of complication is high, such as those with 'stag horn' calculi, uncorrected hypertension or a coagulopathy are best treated in an in-patient setting for proper monitoring for at least 48 hours [3]. Though it is an invasive procedure [6], it is safe and can be used successfully for short- and long term drainage in all children, even in small infants and those with solitary kidneys, and is a valuable adjunct to surgery [7].

CONCLUSION

Once a fused kidney is diagnosed or suspected, further laboratory and imaging evaluation should be performed to assess the status of the kidneys and to look for treatable complications and associated anomalies.

Ultrasound guided percutaneous nephrostomy by direct puncture set is a reliable and cost effective technique for making a temporary drainage pathway urinary obstruction.

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