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Pancake Kidney With Obstructed Moiety: A Rare Renal Fusion Anomaly

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ABSTRACT

Renal fusion abnormalities are rare. Even more rare is pancake kidney. We present a case of a 28-year-old male with symptomatic obstruction of a non-functioning moiety of a pancake kidney. He underwent ureterectomy with a finding of only atretic renal parenchyma at exploration. He recovered well and had resolution of his pain at 3-month follow-up.
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Introduction

Renal fusion anomalies are reported in 0.1–0.2% of all live births. The most extreme form of renal fusion is the pancake kidney (PK), thought to make up less than 10% of all renal fusion anomalies.[1] In this case, both medial poles of each kidney pole become fused, producing an ovoid mass. The pelvis is anteriorly placed and the ureters remain uncrossed. Each collecting system drain its respective half of the kidney and does not communicate with the opposite site.[2] Some case reports were described in the literature, most of them as an incidental finding with conservative management,[3,4] but no case report was found of a PK with a distal ureteral obstruction causing hydroureter and pain requiring surgical intervention.

Case presentation

A 28-year-old male presented to the ED complaining of acute on chronic right lower quadrant pain increasing over the last 4 days, associated with nausea, emesis and dysuria. The patient had a known diagnosis of PK, but was new to our practice. Physical exam revealed normal vital signs tenderness on palpation of the RLQ. There was no CVA tenderness. Initial laboratory work was unremarkable, including urinalysis. A computed tomography scan of the abdomen and pelvis was obtained, revealing a pancake kidney with right hydroureter measuring up to 2.1 cm and a 5 mm calculus at the right UVJ. There was an atretic right/upper moiety draining into the dilated ureter. On the left, there were upper and lower moieties draining into a single, non-dilated ureter (Fig. 1).

Patient was initially taken for cystoscopy with retrograde pyelogram (RP) and ureteral stent placement. Intra-operative findings revealed a very difficult to cannulate right ureteral orifice with approximately 4 cm of distal ureteral obstruction/fibrosis. RP revealed a massively dilated right proximal ureter with suspected right distal ureteral obstruction and no apparent calyces (Fig. 2). It did not appear that the stone was the cause of the

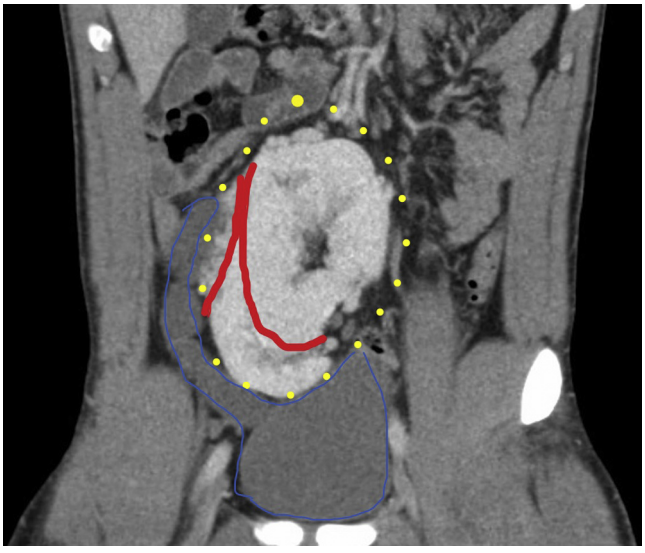

Figure 1. CT Scan revealing ectopic abnormally fused pancake kidney (yellow dots – outline of pancake kidney, blue – outline of obstructed right ureter and bladder, red–separation of renal moieties).

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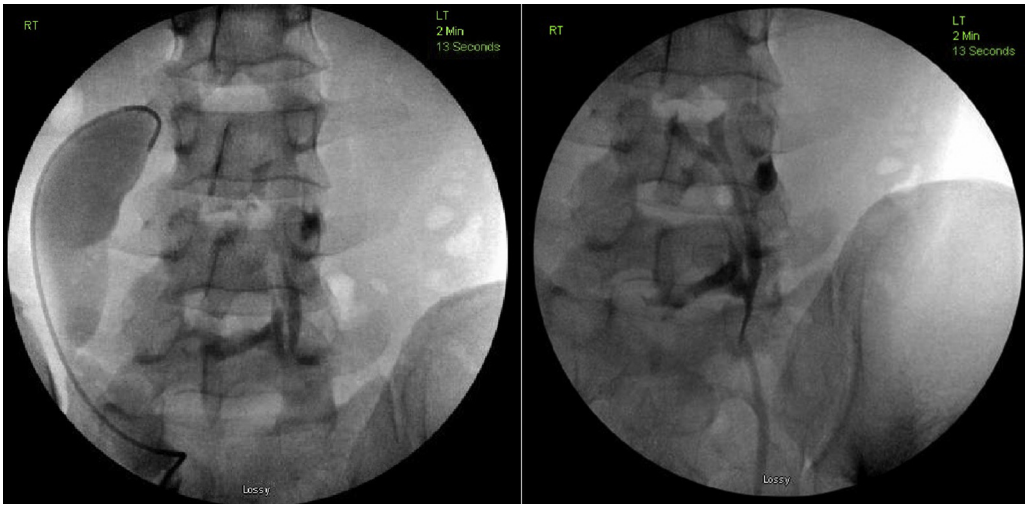

Figure 2. Bilateral retrograde pyelograms revealing a massively dilated right proximal ureter without discernible calyces and left-sided bifid system – medially facing calyces, draining the remainder of the parenchyma.

obstruction. The left RP showed a bifid system, draining the two separate moieties on the left side, consistent with PK (Fig. 2). A double-J ureteral stent was left.

After discussion with patient, we proceeded with exploratory laparotomy, possible ureteral reimplantation, possible resection of right renal moiety. An ectopic, fused kidney in the pelvis with an atrophic right-sided moiety and obstructed, atretic collecting system was found with a clear plane between the right and left moieties, allowing safe excision of the right collecting system (Fig. 3). The remaining atretic parenchyma was cauterized, negating the need for formal partial nephrectomy. A very distended, 15 cm right ureter was also excised (Fig. 3). Because of the obstruction, the distal ureter was left open. Final pathology showed a renal pelvis and ureter with muscular hypertrophy, mural fibrosis, chronic inflammation, and hemorrhage. Drain output showed no evidence of urine leak. Serum creatinine was stable at 0.9 mg/dL. Patient was discharged on post-operative day three. At 3 months of follow-up, the patient had normal renal function and had resolution of his chronic pelvic pain.

Discussion

Wilmer in 1938 was the first to describe the categorization of fusion kidney anomalies, while McDonald and McClellan refined and expanded it in 1957.[5] The most common type of fusion anomaly is the horseshoe kidney, with an incidence of 1 in every 700 autopsies.[6] The exact incidence of PK is unknown, but is assumed to be the rarest type of fused renal ectopic and was first described by Looney in 1926.[7]

This entity is described as a renal mass located in the pelvis which is formed by complete medial fusion of renal parenchyma without intervening septum. Each kidney has its own collecting

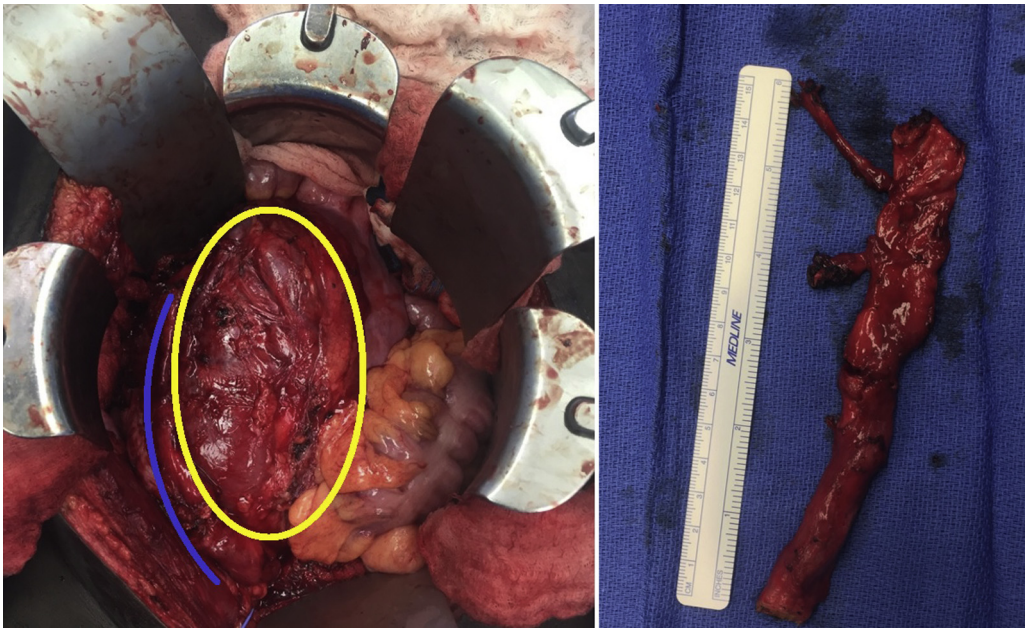

Figure 3. Intra-operative findings of pancake kidney and final pathologic specimen showing atretic calyces, no significant parenchyma and a chronically dilated ureter (yellow line is outline of pancake kidney and blue line is path of dilated right ureter).

system and anteriorly placed short ureters entering the bladder. As with horseshoe kidney, the rotation anomaly of the collecting system and short ureters make them prone to stasis and obstruction. Our patient seemed to have a distal obstruction from fibrosis, possibly from a stone. Our patient also had a bifid left collecting system that seemed to drain two separate moieties.

It is described in the literature that upon surgery of a PK, division of the parenchyma presents potential problems such as renal vascular damage and postoperative renal failure.[8] Careful preoperative scrutiny of the imaging allowed for assessment of the PK anatomy and helped in guiding our dissection during this surgery. Fortunately, in our patient the separation of the two moieties was not necessary and the remaining parenchyma was treated with electrocautery, allowing secure hemostasis as well.

In the literature, most cases describe a conservative management of incidental diagnosed PK [3,4,9] on asymptomatic patients or during other surgical intervention.[10] What makes our case even more interesting is left-sided duplication and the right-sided distal ureteral obstruction resulting in chronic pain that was resolved by the surgical management of this condition.

Conclusion

Pancake kidney is rare and usually asymptomatic. Surgical intervention may be needed occasionally. Careful preoperative scrutiny of imaging can guide surgical therapy. More research is needed to establish the standard management of these patients, which is difficult due its extreme rarity.

Conflict of interest

The authors have no conflict of interest regarding this publication.

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