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FUSED PELVIC KIDNEY DRAINED BY A SINGLE URETER*

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SUMMARY: Tekant G, Bulut M, Akansel G, Yılmaz N. (Department of Pediatric Surgery, Şişli Children's Hospital, İstanbul, Turkey). Fused pelvic kidney drained by a single ureter. Turk J Pediatr 33: 59-63, 1991.

A case of fused pelvic (discoid) kidney drained by a superiorly inserted single ureter is presented. This is the twentieth case of fused pelvic kidney, and the fifth case in which drainage was carried out by a single ureter, to be reported in the English literature. The diagnosis and treatment of this condition is discussed and the relevant literature is reviewed. *Key words: kidney, fused pelvic kidney, discoid.*

Fused pelvic kidney (discoid) is considered to be an extremely rare genitourinary anomaly[1-13]. Goren and Eidelman[13] have recently reported the nineteenth such case. Fused pelvic kidney drained by a single ureter has been reported in four of the documented cases. In this paper, we describe a case of fused pelvic kidney with a superiorly inserted single ureter and review the pertinent literature.

Case Report

A two-year-old boy was admitted to the Pediatric Surgery Clinic of the Şişli Children's Hospital with the chief complaints of abdominal swelling, right lower quadrant pain, and vomiting.

The patient's history was noncontributory. Physical examination revealed a solid, immobile mass in the right lower abdominal cavity which measured 5 × 5 cm in diameter. The blood pressure was 110/70 mmHg. In order to determine the origin of the mass, an emergency abdominal ultrasonography was performed which revealed a cystic hydronephrotic kidney located in the right iliac fossa. The left kidney could not be visualized in its normal anatomical position. The patient was hospitalized for further investigation.

Laboratory studies revealed that the blood urea was 106 mg/dl, creatinine, 2 mg/dl, and blood cell counts and urinalysis were within normal limits. The urine culture remained sterile.

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Plain abdominal radiography showed no significant findings. A voiding cystourethrography (VCUG) demonstrated trabeculation on the right upper portion of the bladder but no vesicoureteral reflux was observed (Fig. 1). Renal isotope scans (Tc 99m-DTPA) demonstrated an ectopic kidney with a single ureter located in the right iliac fossa.

The mass in the right lower quadrant disappeared on the second day of hospitalization, and the blood urea level dropped to 38 mg/dl. A presumptive diagnosis of single ectopic kidney with ureteropelvic junction obstruction was established.

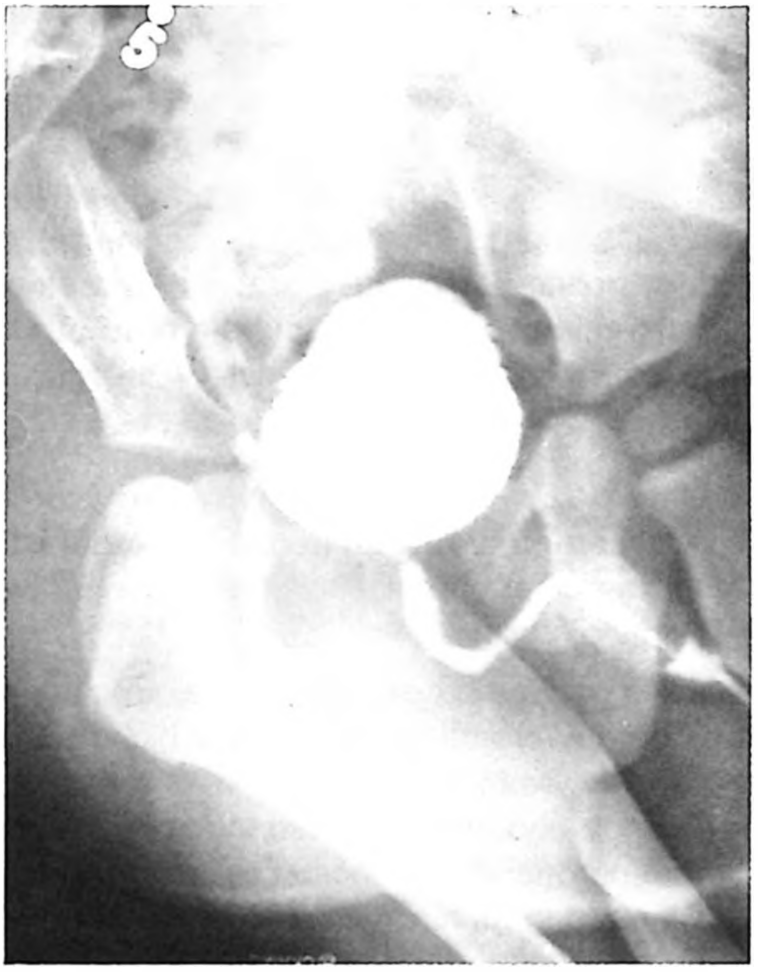

Fig. 1: VCUG demonstrating trabeculation on the right upper portion of the bladder.

To further determine the nature of this urinary abnormality, the abdomen was exposed with a long midline incision extending above and below the umbilicus. The mass was identified as a fused pelvic kidney with a superiorly inserted single ureter (Fig. 2). The renal artery and vein were seen entering the superior pole of the kidney. A dismembered pyeloplasty was performed to correct the ureteropelvic junction obstruction. Further dissection was considered to be hazardous to the

vascular supply of the kidney. The abdomen was closed with a pensore drain placed in the upper angle of the vesical space.

Renal biopsy demonstrated congestion of the glomeruli and tubular changes consisting of cloudy swelling of the tubulus epithelium.

The patient received systemic antibiotics, and had no problem in the postoperative course except for a urine discharge from the drain which was removed on the 13th day. Urinalysis showed no signs of infection, urine cultures remained sterile. Postoperative intravenous pyelography (IVP) demonstrated filtration of contrast material at five minutes and a pelvic fused kidney drained by a single tortous ureter. The patient was discharged on the 15th postoperative day.

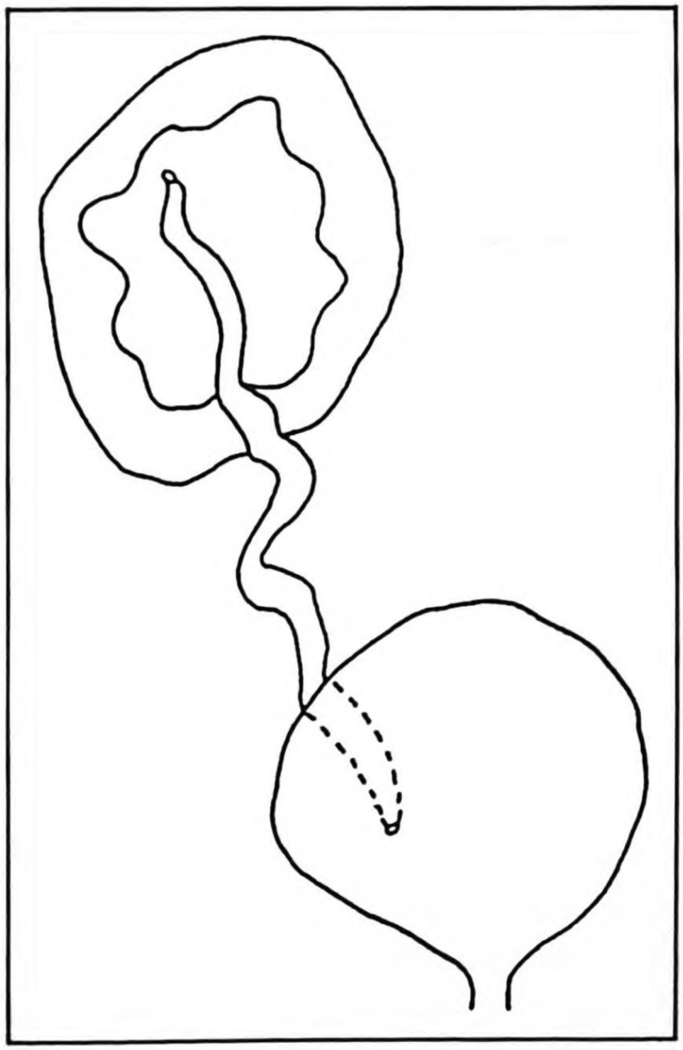

Fig. 2: Schematic drawing of the fused pelvic kidney with a superiorly inserted single ureter.

Discussion

Fused pelvic kidney is a rare congenital urologic abnormality which may be diagnosed in infancy and up until the eighth decade[1-13]. Because of its appearance and shape, this anomaly has been referred to as "cake", "discoid", "shield", "doughnut" or "lump" kidney[4,5,11]. The final shape of the fused kidney is dependent on the time and extent of fusion and on the degree of renal rotation that has occurred. In the lump or cake kidney, extensive joining has taken over a wide margin of renal anlage, so that the total mass is lobulated and both renal pelves drain separate renal areas anteriorly. On the other hand, in the discoid, shield or doughnut kidney, there is more extensive fusion along the entire medial aspect of each kidney, thus providing a better preserved reniform shape with anterior draining uncrossed ureters[11].

Embriologically, two buds of metanephrogenic tissue originate in the 5 mm human embryo in front of the second sacral segment. The kidneys migrate upward to their final lumbar position following movements of ascent, lateral migration, axial deflection and internal rotation. During their ascent in the 30 day nine mm embryo, the umbilical arteries squeeze the nephrogenic blastemas which may lead to fusion. Completely fused kidneys are prevented from ascending and remain ectopic in the pelvis. The weight of the fused kidney is almost equal to that of two separate kidneys. The single ureter draining the fused kidney may be due to regression of the second ureteral bud after fusion of the metanephric blastemas at the ten mm stage. The etiology of the lack of migration and fusion of renal structures is not known[1,3,7,9,11-13].

Vascular supply of the pelvic fused kidney is consistent with its arrested migration. The arterial blood supply is from the aorta near the bifurcation or from the common iliac vessels. Venous drainage is usually into the interior vena cava or into one of the common iliac veins[12,13]. This anomalous blood supply leads to an increased risk of vascular compromise caused by pelvic trauma, vascular disease or pregnancy[7]. In our case, the arterial supply was mainly from the aorta and venous drainage into the inferior vena cava.

It is important to stress that anomalies of the cardiovascular system (tetralogy of Fallot, ventricular septal defect, aortic coarctation), genitourinary tract (testicular maldescent, vaginal agenesis) or gastrointestinal tract (caudal regression, anal anomalies)[1-3,8-12] occur in association with this disorder and that the prognosis depends on these factors. No additional anomalies were found in our case.

Since fused pelvic kidney may be found in all age groups, it may be present with a variety of symptoms such as urine stasis, infection, stone formation, and vascular compromise[1-13]. Fused pelvic kidney does not carry a poor prognosis, however, accompanying complications may result in an unfavorable one. Thus, after the

diagnosis is established, patients should be closely followed for prompt detection of any complications. In our patient the only significant finding was the detection of an abdominal mass, resulting from urinary stasis, which necessitated surgical intervention.

A surgical approach and drainage of the fused pelvic kidney are not recommended because of the variable blood supply[1-13]. However, in our case, surgical intervention was mandatory because of uretero-pelvic junction obstruction of the single ureter. No further exploration or dissection has been performed.

This patient is the twentieth case of fused pelvic kidney, and the fifth case in which drainage was carried out by a single ureter, to be reported in the English literature. During the eight month follow-up, the patient remained clinically healthy, and laboratory and radiologic studies for renal functions were within normal limits.

In conclusion, the fused pelvic kidney with its presentation, diagnosis and prognosis remains a challange to the physician.

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