

SOLITARY PELVIC KIDNEY: AN INCIDENTAL CASE

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We report a case of solitary pelvic kidney. This uncommon condition was detected incidentally and diagnosed with intravenous pyelography (IVP), computed tomography (CT), and retrograde pyelography (RP).

Key words: Pelvic kidney, Renal Agenesis

INTRODUCTION

When the mature kidney fails to reach its normal location in the renal "fossa", the condition is known as renal ectopia. This classification of ectopia is based on the position of the kidney within the retroperitoneum. The pelvic kidney is located opposite the sacrum and below the aortic bifurcation. When a pelvic kidney is found, the contralateral kidney is frequently normal, but sometimes there may be congenital defects associated with it¹⁾ Although the incidence of pelvic kidney in autopsy series is not so rare, i.e., one in 2,100 to 3,000 autopsies, solitary pelvic kidney is rare indeed, i.e., one in far more than 22,000 autopsies²⁾. Recently however, urodiagnostic visualization has made more frequent diagnosis possible. As a result, we could incidentally find a case with this uncommon anomaly.

CASE REPORT

A 15-year-old girl was referred to our clinic for evaluation of abnormal IVP findings obtained incidentally during medical treatment for hyperuricemia and hyperthyroidism at another clinic. She had a high fever attack that lasted several days three years previously, but had no other history suggesting urological disorders. She appeared to be well developed and healthy in appearance and showed normal intelligence. Her height was 155 cm and she weighed 47 kg. Her blood pressure

was 120/80 mmHg. Menarche had not started yet. The development of her external genitalia was normal but there was no pubic hair. The vagina was short and blind. Skeleton abnormalities consisting of protrusion of the right 2nd rib and moderate lumbar scoliosis were detected by bone X-ray. The urinalysis results were normal and, except for serum uric acid and thyroid function, all of the laboratory data including serum creatinine level were within normal limits. The patient's karyotype was 46,XX.

However, the left kidney could not be visualized with IVP performed at the other clinic. The right kidney was ectopic showing a hydronephrotic pelvic kidney with incomplete rotation, but we could not detect the left kidney in either the renal "fossa" or the pelvic cavity even CT scan. The right pelvic kidney was seen as a hydronephrotic renal pelvis with calyces anterior to the parenchyma and fetal lobulation or a pancake-like shape in the parenchyma (Fig. 1, 2). In addition, neither the uterus nor definite gonads could be identified by either CT scan or ultrasonography. Cystoscopically, the trigone and the bilateral ureteral orifices were normal, while retrograde pyelography performed at the same time showed hydronephrosis in the right kidney with high insertion of the ureter into the pelvis accompanied by junctional stenosis. Furthermore, the left ureter was blind (Fig. 3), although no vesicoureteral reflux was seen. Pyelo-

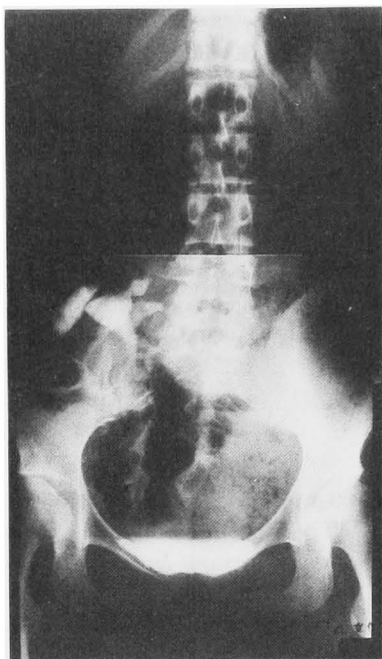


Fig. 1. IVP: The left kidney could not be visualized. The right pelvic kidney with hydronephrosis and incomplete rotation is shown.

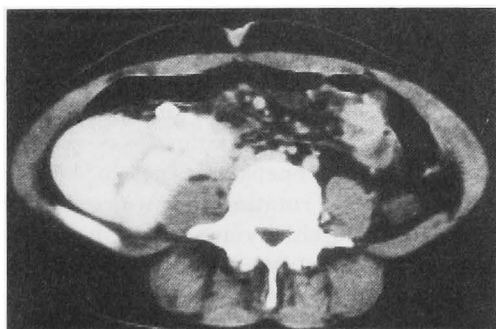


Fig. 2. Enhanced CT: Fetal lobulation or pancake-like shapes in the parenchyma of the right kidney in the pelvic cavity. Ectatic renal pelvis and calyces anterior to the parenchyma.

plasty was recommended but refused.

DISCUSSION

A kidney ectopically located in the pelvis fails to follow the normal course of embryonic ascent from its early position as a metanephric blastema posterior and lateral to the urogenital sinus, to its

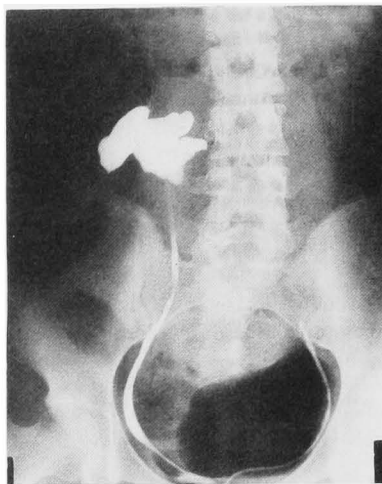


Fig. 3. Retrograde pyelography: Hydronephrosis of the pelvic kidney due to high insertion of the right ureter into the pelvis accompanied by junctional stenosis. The left ureter is blind.

cranial position high in the flank. Development of the parenchyma is completed during ascent while both the metanephrogenic mass cranially and the ureter caudally undergo complex maneuvers of rotation and migration. The final position of the kidney and ureter often reflects the stage of embryonic development of both structures when migration ceases¹⁾. Although the existence of abnormal intraparenchymal induction of the mesenchyme into the glomerular structures and of the ureteral bud epithelium into the collecting tubules is not always seen, it is not uncommon to see persistently aberrant blood vessels, external pelvis, fetal lobulation, pancake or discoid shapes in the parenchyma, and short ureters with complex rotational anomalies of completely crossed renal ectopia^{3,4)}. These abnormalities alone may not lead to illness. However, as demonstrated in the literature and the present case, a high incidence of genitourinary and skeletal anomalies accompanies pelvic kidney^{1,5)}. In cases with pelvic kidney or renal agenesis, genital anomalies arising from the mal-development of the mesonephric or paramesonephric duct have been reported^{1,6,7)}. In the present case, the uterus could not be detected and the vagina

was short and blind. The mesonephros and its duct may act as an inducer for metanephric development as well as ascent²⁾. If the mesonephros is absent or abnormal, then the metanephros or final kidney may fail either to develop or to ascend depending upon the time of occurrence of the mesonephric abnormality.

Furthermore, the mesonephric duct may be necessary for the proper development of the Müllerian or paramesonephric duct. In other words, the mesonephros may be required for metanephric development and ascent, while the mesonephric (Wolffian) duct may be required for paramesonephric (Müllerian) duct development. If mesonephros and Wolffian duct resorption occurs early in the female, then absence of the kidney and Müllerian duct agenesis will occur. If it occurs later, i.e., after the ureteral bud joins the metanephros, but before the Müllerian duct development is complete, then the kidney will assume a pelvic position and lower Müllerian duct agenesis, i.e., vaginal atresia, will occur. The precise embryological interpretation of the solitary pelvic kidney in our case is difficult but it could be attributable to the resorption of the mesonephros on both sides; one side occurred early and the other a little later but before the Müllerian duct development was completed.

Besides genital anomalies arising from incomplete development of the Wolffian or Müllerian duct, a high incidence of musculoskeletal, gastrointestinal, cardiac and central nervous system anomalies has been reported in patients with pelvic kidney or renal agenesis^{1,6,7)}. In patients with solitary pelvic kidney, the increased risk of renal failure in the second or third decade of life due to recurrent infection

from vesicoureteral reflux, hydronephrosis and calculi makes early diagnosis and critical longitudinal care mandatory^{8,9)}. Furthermore, recent surgical techniques have markedly decreased the morbidity and mortality rates, so that aggressive surgical therapy is recommended in such cases of urinary malformation as the solitary pelvic kidney presented here.

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和文抄録

片側腎欠損を伴う骨盤腎：偶然発見された1例

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片側腎欠損を伴う骨盤腎を偶然発見し，排泄性腎盂造影，CT および逆行性腎盂造影にて診断した。

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