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Fused Cake Kidney Combined with Hypoplastic Thumb : A Case Report[1]

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During embryologic development, many renal anomalies, including fusion and ectopia, can occur. Among them, fused cake kidney is a rare developmental anomaly. We report a case in which this condition was combined with hypoplastic thumb. Ultrasonographic, scintigraphic, CT and MRI findings of this rare condition are presented.

Index Words : Children, genitourinary system
Kidney, abnormalities
Kidney, MR

Complicated renal embryologic development can result in many renal anomalies, including fusion and ectopia, and these two conditions may be associated with anomalies of other organs outside the urinary tract. We report a case of fused pelvic kidney combined with hypoplastic thumb, and the related findings of ultrasonography, renal scintigraphy, CT and MR imaging.

Case Report

A newborn male with hypoplastic left thumb was examined to determine whether associated anormalies were also present. He was born at term(40 weeks) by spontaneous vaginal delivery, and at birth weighed 3. 2kg. He had no sibling. During the pregnancy, his mother had experienced no medical problem.

Ultrasonography showed an ovoid, mixed echogenic mass, with the appearance of a lotus root, in the pelvic cavity (Fig. 1). The kidney was not identified on either side of the flank. Tc-99m DMSA renal scintigraphy revealed an ovoid, lobulated radioactive mass just above the urinary bladder in the midline (Fig. 2), and a CT scan of the pelvis showed an ovoid, reniform, well-enhanced solid mass in the pelvic cavity (Fig. 3). T1-weighted coronal MR imaging also showed a round, well-defined, lotus root-like solid mass in the same location, representing a fused cake kidney, with a well differentiated renal cortex and medulla (Fig. 4). On plain chest radiography, a focal hyperlucent area was noted in the right lower lobe.

Two months later, due to the sudden onset of coughing and dyspnea for one day, the patient was admitted to the hospital. The chest CT scan revealed an area of hyperlucency in the right lower lobe, suggesting congenital lobar emphysema. Multifocal patchy pneumonic consolidations were present in both lungs (Fig. 5).

On the 45th hospital day, severe hypoxia developed and he died of respiratory failure. An autopsy was not performed.

Discussion

Fused pelvic kidney, fused cake kidney, or lump kidney is defined as an anomaly in which the entire renal substance is fused into one mass lying in the pelvis, and gives rise to two seperate ureters that enter the bladder in the normal way(1). The two kidneys are at the same level and vertically oriented, and their pelves may face each other (unilateral disc or cake kidney) or face forward(unilateral lump kidney) (2).

Fused cake kidney is a rare anomaly. In 2,153 autopsies in children, Rubinstein and co-workers found 180 instances of urinary tract anomaly, none of which represented a complete fusion of the kidneys(3).

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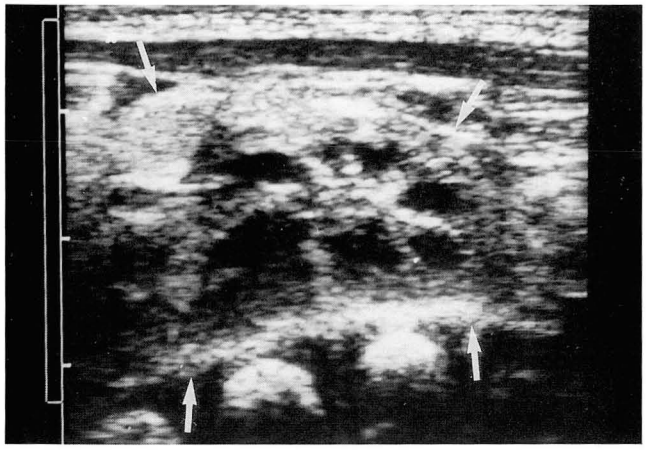


Fig. 1. Ultrasonogram of the pelvis shows a well defined, lotus root appearing mass lesion(arrows) in the pelvic cavity.

While the mesonephric stage continues, the developing permanent kidneys arise caudally from the ipsilateral nephrogenic cord as two condensed masses of mesoderm. At the same time, the ureteral buds arise from each wolffian duct. During the ascent to their final position, where they meet the suprarenals at approximately 8 weeks, the kidneys establish temporary vascular connections; first with the middle sacral arteries, then with the iliac arteries, and finally, with the aorta. At the same time they rotate about their axes through 90 degrees so that the dorsal border becomes lateral and the hilus lies medially (4). Fusion occurs when each metanephric mass is still in the pelvis early in its ascent. An abnormally placed umbilical artery may force the two metanephric masses into opposition, leading to fusion. Following this, ascent to the normal position is impaired by the retroperitoneal structure.

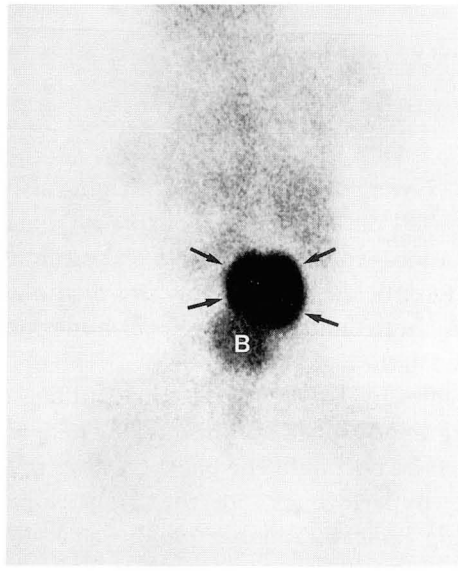


Fig. 2. Renal scintigram with Tc99m DMSA reveals an ovoid, lobulated radioactive mass (arrows) just above the urinary bladder(B) in the midline.

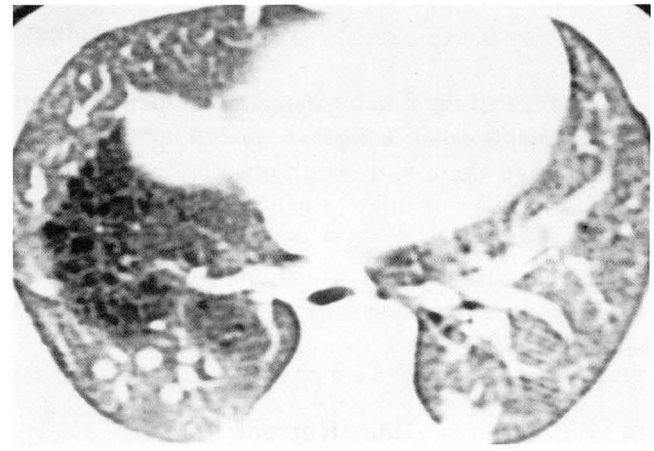
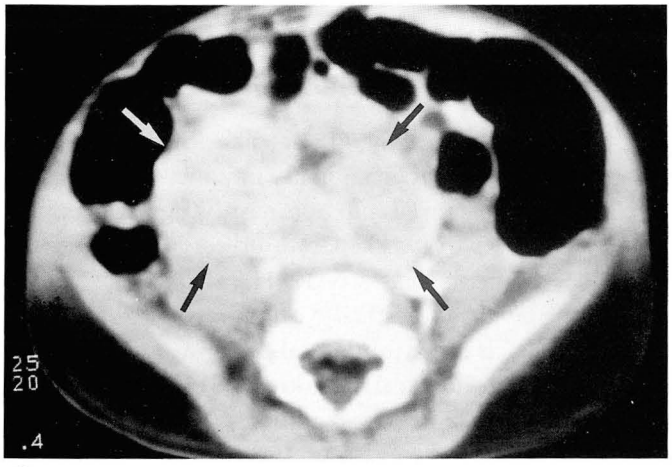


Fig. 5. Two months later, after onset of sudden coughing and dyspnea, the chest CT scan (lung window setting) shows an area of hyperlucency in the right lower lobe, basally, with multifocal pneumonic consolidations in both lower lobes.



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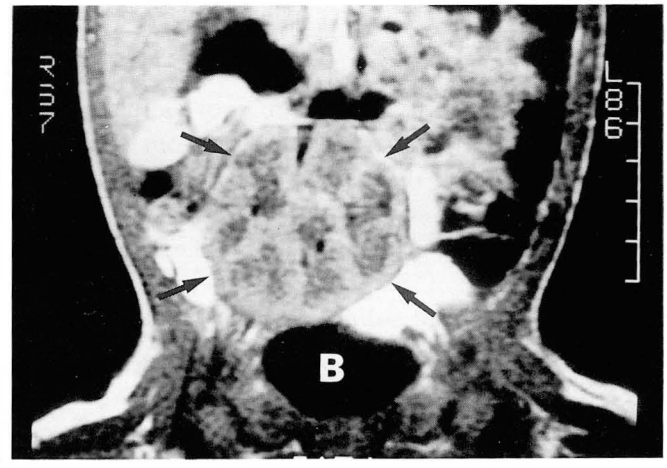

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Fig. 3 & **4.** Contrast-enhanced CT scan and T1 weighted coronal MR image show typical findings of fused cake kidney (arrows) in the pelvic cavity just above the urinary bladder(B).

When the kidney stays in the pelvis, its blood supply is usually derived from the terminal aorta or the iliac artieries. Grossly, the kidney has a flat smooth posterior surface and a lobulated anterior surface, histologically, the fused pelvic kidney shows immature glomeruli, cystic change, and enlarged dilated tubules (5).

Renal ectopia and fusion may be associated with anomalies of many other organs, the most common of which are found in the skeletal system, and include asymmetry of the skull and ribs, or vertebral anomalies and an absence of bones (4). Associated anomalies in the cardiovascular and gastrointestinal tract are not uncommon (4). In our case, fused cake kidney was associated with hypoplastic thumb and probable congenital lobar emphysema in the right lower lobe.

Ectopic and fused kidneys may be complicated by ureteropelvic junction obstruction because of malrotation, abnormal renal shape and aberrant vasculature (2). Vesicoureteral reflux is a frequent problem, due to the tortuous and short ureters of ectopic and fused kidneys, such kidneys may be asymptomatic and be found incidentally as an abdominal mass (4). In pelvic surgery, such as cystectomy for bladder malignancy in the presence of fused pelvic kidney, the preservation of aberrant renal vasculature when performing lymph node dissection requires great care (5).

We report a case of fused cake kidney associated with hypoplastic left thumb and probable congenital lobar emphysema in the right lower lobe, and its typical findings on ultrasonography, scintigraphy, CT and MR imaging.

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모지형성부전과 동반된 골반내 융합이소신장:1예 보고[1]

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김기준 · 변재영 · 김학희 · 신경섭

복잡한 신장의 발생과정 중 융합 및 이소성을 포함한 다양한 신장의 선천성 기형이 생길 수 있으며 그 중 골반내 융합 이소신장은 매우 드문 선천성 기형으로 알려져있다. 또한 신장의 융합 및 이소성 기형은 다른 기관에 선천성 기형을 동반할 수 있다. 저자들은 초음파, 방사선동위원소 신장스캔, 전산화단층촬영 및 자기공명영상으로 진단된 모지형성부전과 동반된 골반내 융합이소신장 1예를 보고하는 바이다.

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14 : 30 − 15 : 00	Advance in Abdominal MRI	최병인 교수 (서 울 대)
15 : 00 − 15 : 30	Advance in Musculoskeletal MRI	서진석 교수 (연 세 대)
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16 : 40 − 17 : 30	MR Spectroscopic Imaging : Basics & Clinical Applications	Dr. Peter B. Barker (Johns Hopkins University)

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