Original Research Article

Problem of fused kidneys - Our observations

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Abstract

Genitourinary tract tops the list of developmental anomalies – 30-40% of malformed individuals. These developmental anomalies comprise a diversity of abnormalities ranging from complete absence to aberrant location, orientation, shape, form, fusion, number and vascular attachments. Amongst these wide range of anomalies, fusion anomalies are not uncommon. Horseshoe kidney is the commonest fusion anomaly which accounts for about 0.25% of the population. Unilateral fused kidney with inferior ectopia is the next most common fusion anomaly with an incidence of 1:1000. The other fusion anomalies are Sigmoid or S – shaped kidney, Lump kidney, L shaped kidney, Disc or doughnut kidney and Unilateral fused kidney with superior ectopia. These fusion anomalies are clinically important because they are malrotated, abnormally placed with anomalous blood supply. They are predisposed to various complications like hydronephrosis, infection, urolithiasis and neoplasm. This is a study of 10 patients who presented with fusion anomalies, in the department of Osmania Hospital, Afzalgunj during the period of 2013-2015. The clinical presentations, modalities of arriving at diagnosis, associated complications and the management are discussed.

Key words

Fused kidneys, Developmental anomalies, Complications.

Introduction

Developmental anomalies are highest in the genitourinary tract (30-40%) [1]. Of the numerous anomalies of the kidney like number, volume, structure, form, rotation and vascular attachments, fusion anomalies are not uncommon.

Fusion anomalies are clinically important conditions because they are predisposed to

various complications like hydronephrosis, urolithiasis, infection and neoplasia.

The fused kidneys are abnormally placed, malrotated and the vascular supply is generally anomalous.

Types of fusion anomalies

Horseshoe kidney

It is the most common of all renal fusion anomalies [2]. The anomaly consists of two distinct renal masses lying vertically on either side of midline connected at their respective lower poles by parenchymatous or fibrous isthmus that crosses the midline of the body.

History

In 1564, Botallo provided the first description and illustration of Horseshoe kidney. In 1820, Morgangi described the first diseased horseshoe kidney.

Incidence

Horseshoe Kidney occurs in 0.25% of the population or about 1:400. This abnormality has been discovered in all age groups. It is found more commonly in males by a 2 to 1margin. No definite genetic predisposition has been identified. It has been reported in identical twins and siblings.

Embryology

The fusion abnormality occurs between 4^{th} and 6^{th} week of gestation, after the ureteral bud had entered the renal blastema. It is thought that metanephric masses that lie too close to midline, abnormal positioning of the umbilical or common iliac arteries or anomalous formation of the tail of the embryo predispose to this fusion.

Ascent of the Horseshoe kidney is arrested when the isthmus or fused segment reaches the inferior mesenteric artery. The fusion also prevents normal medial rotation of the kidneys so that the pelvis are directed anteriorly and the ureters overlie the isthmus.

Description

In 95% of cases, the kidneys join at lower poles. Generally the isthmus consists of parenchymatous tissue with its own blood supply and occasionally it is a flimsy fibrous band. It is located at the L3 or L4 vertebrae just below the junction of inferior mesentric artery with aorta, anterior to the aorta and inferior vena cava.

The calyces are normal in number, but point medially. The axis of the pelvis remains in vertical or obliquely lateral plane on a line drawn from lower to upper poles. The lower most calyces extend medially to drain the isthmus and may overlie the vertebral column.

The ureter may insert high on the pelvis and lie laterally. They cross over the isthmus. The lower ureters usually enter the bladder normally and rarely are ectopic.

Blood supply

In 30% of cases it consists of one renal artery to each kidney. The isthmus and adjacent parenchymal masses may receive a branch from each main renal artery of they may have their own arterial supply from aorta originating either above or below the isthmus or may be supplied by branches form inferior mesenteric, common iliac or external iliac or sacral arteries.

Associated anomalies

The organ systems most commonly involved include skeletal (scoliosis, myelodysplasia), cardiovascular (ventriculoseptal defects), Gastrointestinal (imperforate anus) and central nervous system (neural tube defects) [3].

It is found in 20% of patients with trisomy 18 and 60% of females with Turner's syndrome. Hypospadias and undescended testes are seen in 4% of males. Bicornuate uterus or septate vagina is seen 7% of females. Duplication of ureter is seen in 10% of patients with Horseshoe kidney. Vesicoureteric reflux is noted in more than half of the individuals.

Clinical presentation

One third of patients remain asymptomatic. When symptoms are present they are related to hydronephrosis, infection or calculus formation. An abdominal mass is palpated in 5-10% of individuals.

Complications

- PUJ Obstruction: PUJ Obstruction causing hydronephrosis is seen in 1/3rd of patients with Horseshoe kidney. High insertion of ureter, its anomalous course over isthmus and anomalous blood supply contribute to this obstruction.
- Renal Calculi: Renal Calculi have been noted in 20-80% of individuals with Horseshoekidney [4].
- U.T.I: Signs and symptoms of urinary tract infections occur in 30% of individuals with Horseshoe kidney.
- Renal Neoplasms: Wilm's tumor is 8 times more common. Renal cell carcinoma and transitional cell carcinoma of renal pelvis are also more common. The cancers are commonly seen arising from isthmus.

Diagnosis

In 33% of patients it is an incidental finding during investigation of other disorder. It is not suspected until an Ultrasonogram or I. V. U has been obtained. The characteristic findings on I. V. U are:

- Low lying kidneys
- Vertical renal axis
- Orientation of collecting systems which is directed posterior to each renal pelvis with the lowermost calyx pointing caudally or even medially.
- High insertion of ureter into renal pelvis and anteriorly displaced ureter.

Rarely R.G.P or CT scan may be required to confirm the diagnosis.

Management

Surgical approach

The appropriate surgical approach to the Horseshoe kidney is directed by the nature of the problem that requires correction, size and body habitus of the patient and the surgeon's preference [5].

a. Anterior subcostal incision:
   It provides satisfactory extraperitoneal exposure to one side of Horseshoe kidney. The insicion is extended medially if vascular control of isthmus is required.
b. Transverse abdominal incision:
   It provides satisfactory exposure for all operative procedures.
c. Midline abdominal transperitoneal approach:
   It also provides satisfactory exposure for all operative procedure.

Treatment of puj obstruction

Foley Y-V plasty is well suited because of high insertion of ureter into renal pelvis. Dismembered Anderson Hynes pyeloplasty is recommended when renal pelvis is extremely redundant [6, 7].

Treatment of renal calculi

Both PCNL and ESWL [8] can be used apart from open surgery.

Nephrectomy for tumor [6]

An arteriogram is always need to provide a road map of vascular supply. The involved kidney, adrenal gland and atleast half of the isthmus and investing Gerota's fascia are removed enbloc.

Division of the isthmus

No Patient benefitted from division of the isthmus for relief of pain. This idea has now been largely repudiated [9, 10]

Crossed fused ectopia

Crossed fused ectopia is the second most common fusion anomaly. 90% of crossed fused ectopic kidneys are fused to their ipsilateral mate.

Types of fusion anomalies [11]

- Unilateral fused kidney with inferior ectopia [10]

- Sigmoid or S shaped kidney
- Lump or cake kidney
- L shaped or tandem kidney
- Disc, shield or doughnut kidney
- Unilateral fused kidney with superior ectopia

Incidence

The incidence is 1:1000. Unilateral fused kidney with inferior ectopia is the most common anomaly. Sigmoid or S shaped kidney is the second most common anomaly. Fusion with superior ectopia is the least common anomaly.

Left to right ectopia is seen three times more frequently that right to left. The male female ratio is 2:1 [12].

Embryology

The factors responsible for change in kidney position during gestation is still undetermined. Fusion may occur when the renal masses are still in true pelvis or during later stage of ascent. Following fusion, ascent to normal position is impeded by midline retroperitoneal structure.

The final shape of the fused kidney depends upon the time and extent of fusion and the degree of renal rotation that has occurred. No further rotation is likely once kidneys have joined.

In every case of crossed ectopia, the ureter from normal kidney enters the bladder on the same side, whereas that of ectopic kidney crosses the midline at the pelvis brim and enters bladder on the contralateral side.

Description

Inferior ectopia

Of all unilaterally fused kidneys, 2/3 involve inferior ectopia. The upper pole of the crossed kidney is attached to the inferior aspect of the normally placed kidney. Both pelvis are placed anteriorly.

Sigmoid or S - shaped kidney

The crossed kidney is inferior with the two kidneys fused at adjacent poles. Fusion occurs after complete rotation so that each pelvis is oriented correctly and faces opposite direction.

Lump kidney

The total kidney mass is irregular and lobulated due to extensive fusion. Ascent occurs as far as the sacral promontory. Both renal pelvis are anterior and drain separate areas of renal parenchyma. The ureters do not cross.

L shaped or tandem kidney

Crossed kidney assumes a transverse position at the time of fusion and lies in the midline or contralateral paramedian space anterior to the L4 vertebra. The ureter from each kidney enters the bladder on its respective position.

Disc, shield, doughnut or pancake kidney

Kidneys are fused at the medial borders along its entire length. The lateral aspect of each kidney retains its normal contour .The pelvis are anteriorly placed and ureters are uncrossed. Each collecting system drains its respective half of the kidney and does not communicate with the opposite side.

Superior ectopic kidney

The crossed kidney lies above the normal kidney and the lower pole of crossed kidney fuses with the upper pole of uncrossed kidney. Both pelvis lie anteriorly.

Blood supply

The crossed ectopic kidney is supplied by one or more branches from aorta or common iliac artery. The normal kidney frequently has an anomalous blood supply with multiple renal arteries originating from aorta at various levels.

Associated anomalies

The highest incidence of associated anomalies are seen in solitary crossed renal ectopia but they are low in crossed renal ectopia .

The most frequent are:-

Imperforate anus - 4%

Orthpaedic anomalies - 4%

Skeletal anomalies
Septal cardiovascular anomalies

Most orthotopic renal units are normal abnormality is commonly seen in crossed kidney and consist of PUJ Obstruction, cystic dysplasia and carcinoma.

Clinical presentation

Majority of patients are asymptomatic. Defects are discovered incidentally. The abnormal position and anomalous blood supply impede urinary drainage and cause hydronephrosis, urinary tract infections and renal calculi. Patients present with abdominal pain, pyuria, hematuria and urinary tract infections.

Diagnosis

Fusion anomalies are diagnosed by ultrasound, I.V.U. and radionuclide scans.

Management

Surgery is indicated for complications like obstruction, calculi [7] and malignancy. Renal angiography is a requirement prior to extensive surgery on these ectopic kidneys because of anomalous blood supply. The operative techniques for horseshoe kidney are applicable to crossed fused kidneys also.

Prognosis

Patients will have normal longevity and prognosis except for increased risk of obstruction, infection and calculus formation.

Materials and methods

During the period of 2 years (2013-15) from around 1160 urological admissions in Osmania Hospital, we have studied 10 cases of fused kidneys. 7 of them were Horseshoe kidneys. 2 were unilateral fused kidneys with inferior ectopia (one from left to right and another from right to left) and there was one lump kidney.

Age of the patients ranged from 8 years to 65 years. Most of the patients were seen in the 20-40 years age group. Among the ten, there were 5 male and 5 female patients.

Results

Total no. of cases - 10

Horseshoe Kidney - 7
Crossed fused ectopia - 2
Lump Kidney - 1

Age incidence

Less than 20 years - 1
20-40 years - 7
More than 40 years - 2
Males: Females = 5:5 (1:1)

Clinical presentation

Most common presentation was pain abdomen, 3 patients gave history of recurrent U.T.I. A lump was palpable in 3 patients, hypertension was noted in 1 patient

Abdominal pain - 8
Recurrent U.T.I. - 3
Palpable lump - 3
Hypertension – 1

Investigations

The routine basic workup of these cases included urine analysis, urine for C/S, haematological and biochemical study and plain X-ray KUB. X-ray KUB revealed calculus disease in 8 patients.

U.S.G. and I.V.U. were done in all cases which revealed the fusion anomaly. RGP was done in two cases of crossed fused ectopic kidney and one case of lump kidney to confirm the diagnosis. Radionuclide renal scan was done in one patient who presented with lump kidney.

Complications

Urolithiasis is the most common complication noted in our series. It was seen in 8 out of 10 patients. Bilateral renal calculi were seen in one patient. Calculus pyonephrosis was noted in 2 patients. One patient with right crossed fused ectopic kidney had renovascular hypertension.

Calculus - 6
Calculus Pyonephrosis - 2
Renovascular hypertension - 1
Neoplasia - Nil

Management

Pyelolithotomy was done in 6 patients and Heminephrectomy was done in 2 patients who presented with calculus pyonephrosis. The patient who presented with lump kidney with recurrent U.T.I. and right crossed fused ectopic kidney with Hypertension were treated conservatively.

Pyelolithotomy - 6
Heminephrectomy - 2
Heminephrectomy - 2

Discussion

Developmental anomalies are highest in the genitourinary tract, in about 30-40% of malformed individuals.

Of the numerous congenital anomalies of the kidney like number, volume, structure, form, rotation and vascular attachments, fusion anomalies are not uncommon.

Horseshoe kidney is the commonest fusion anomaly which accounts for about 0.25% of the population. In our series it is seen in 7 out of 10 cases. Unilateral fused kidney with inferior ectopia is the next most common fusion anomaly with an incidence of 1:1000.Unilateral fused kidney with inferior ectopia is seen in 2 of our patients. The other fusion anomalies are S or Sigmoid kidney, Lump or cake kidney or tandem kidney, Disc or doughnut kidney and Unilateral fused kidney with superior ectopia. In our series, there is one lump kidney.

These fusion anomalies are clinically important because they are abnormally placed, malrotated with anomalous blood supply. They are also predisposed to various complications like hydronephrosis, infection, urolithiasis and Neoplasma.

Most of the patients present with vague symptom like abdomen and dysuria. In our series pain abdomen and recurrent UTI are the most common presentation.

Among the investigations, IVU is the best investigation to detect fusion anomalies. Other investigations like Retrograde Pyelography and Radionuclide scan are required to confirm the diagnosis.

Among the various complications the fusion anomalies areprone for, urolithiasis is the most common complication noted in our series. It is seen in 6 out of 8 cases. 2 cases presented with calculus pyonephrosis. Renovascular hypertension is seen in one patient with crossed fused ectopic kidney due to anomalous blood supply.

Conservative treatment like antibiotics and analgesics relieve the symptoms in the absence of complications. It has been shown that division of isthmus and nephropexy do not relieve the pain [11, 12]. Division of Isthmus and Nephropexy are not done in any of our cases.

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