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Unilateral renal agenesis with subseptate uterus and sacrococcygeal teratoma: A unique triad

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Abstract

Unilateral renal agenesis (URA) is a rare condition with a reported incidence of 0.93–1.8 per 1000 autopsies. It is commonly diagnosed as an incidental finding on imaging.URA is frequently associated with other genitourinary anomalies. Different associations have been described in both males and females, however, to our knowledge, it has not been reported with subseptate uterus (SSU) and sacrococcygeal teratoma (SCT) in the same individual. Here, we present a unique case of URA with SSU and SCT.

Key words: Sacrococcygeal teratoma; septate uterus; subseptate uterus; renal agenesis

Introduction

Congenital unilateral renal agenesis (URA) is rare and occurs in 0.93–1.8 per 1000 autopsies.[1]

It is commonly diagnosed as an incidental finding on imaging. It is frequently associated with other genitourinary anomalies in both males and females. Genital anomalies are more common in females and constitute approximately 37–60%. Among various genital anomalies, subseptate uterus (SSU) is not commonly seen with URA. Approximately 50% of the patients with solitary absent kidney have associated urological anomalies along with skeletal, gastrointestinal, and cardiovascular abnormalities.[2] URA has not been previously reported with sacrococcygeal teratoma (SCT). SCT is the most common germ cell tumor in newborns, occurring in 1 in 40000 live births with a marked female predominance (75%).[3] Here, we present a unique case of URA associated with SSU and SCT.

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Case Presentation

A 1½-year-old female child presented in our radiology department with a palpable mass on the lower back with nocturnal enuresis and recurrent urinary tract infections (UTI). On examination, a large mass was present on the lower back. Initially, ultrasound was performed that demonstrated a large heterogeneous lesion with multiple areas of cystic necrosis on the lower back. A small uterus was present showing dilated central cavity [Figure 1]. Then, computed tomography (CT) of the abdomen and pelvis was done that demonstrated a large heterogeneously enhancing soft tissue density mass centered in the presacral space measuring approximately 12.1 × 9.0 × 7.5 cm [Figure 2A and B]. The mass was arising from the distal sacrum and coccyx with extension into the sacral spinal canal. It was compressing and displacing the rectosigmoid colon, uterine cavity, and urinary

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bladder anteriorly and showing infiltration in to adjacent gluteal muscles. The uterus showed dilated fluid-filled endometrial cavity with a midline partial septum dividing the endometrial cavity. The septum was not extending up to endocervical canal and represents a SSU [Figure 3]. Right kidney was not present in right renal bed and also not seen elsewhere in the abdomen [Figure 4]. Left kidney was enlarged showing compensatory hypertrophy. Laboratory findings were remarkable for raised serum levels of alpha-fetoprotein. Biopsy of the mass was done and histopathological findings confirmed SCT of yolk sac variety.

Discussion

URA is generally thought to result either from failure of the development of the ureteric bud or to a defect of its interaction with the metanephric blastema involving one side of the urinary tract.[4] Left kidney is more frequently involved than the right one with male-to-female ratio of 1.8.[2]

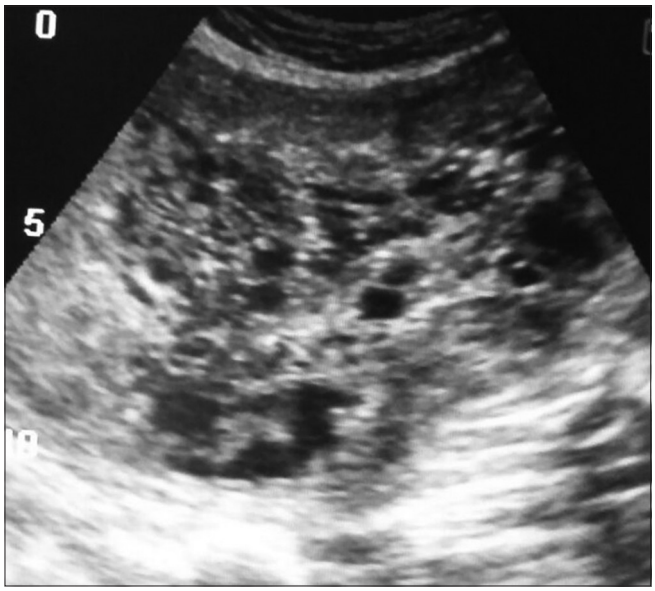

Figure 1: Ultrasound image showing a large heterogeneous lesion with multiple areas of cystic necrosis at lower back

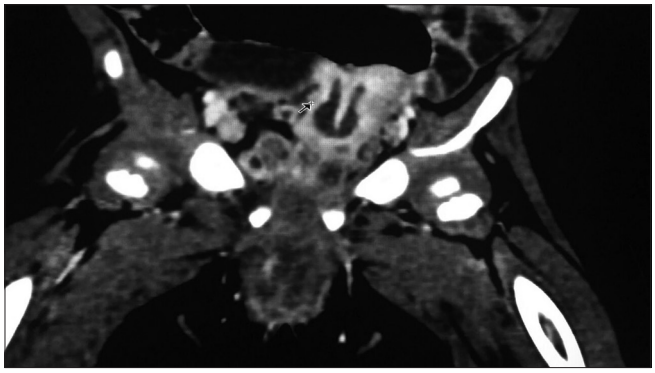

Figure 3: Coronal section of contrast-enhanced CT scan abdomen demonstrating SSU associated with SCT

However, in this case, right kidney was absent. It is frequently associated with other genitourinary anomalies in both males and females. Approximately 50% of the patients with solitary absent kidney have associated urological anomalies such as absent ureter, vesicoureteral reflux, ureterovesical and ureteropelvic junction obstruction. Similarly, skeletal, gastrointestinal, and cardiovascular abnormalities have also been reported.[2] URA may also present as a part of multiorgan syndrome. Mayer–Rokitanski–Kuster–Hauser syndrome is a combination of vaginal agenesis, abnormal uterus, renal, and skeletal anomalies. Similarly, URA has also been described with Herlyn Werner Wunderlich syndrome, Kallmann syndrome, Brachio-oto-renal syndrome, trisomies and Turner syndromes.

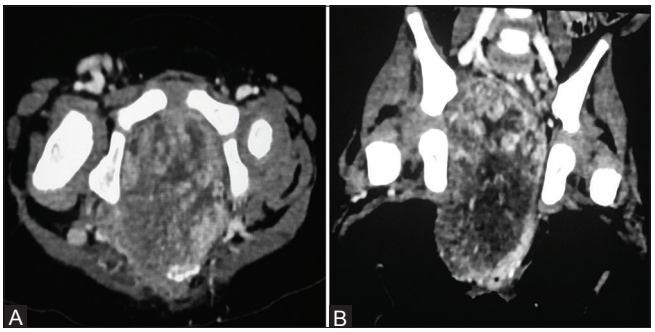

Figure 2 (A and B): (A) Axial image of contrast-enhanced CT scan abdomen showing a large heterogeneously enhancing soft tissue tumor mass lesion involving presacral space with areas of cystic necrosis in it. Biopsy proven SCT. (B) Coronal image of contrast-enhanced CT scan abdomen showing a large heterogeneously enhancing soft tissue tumor mass lesion involving presacral space with areas of cystic necrosis in it. Biopsy proven SCT

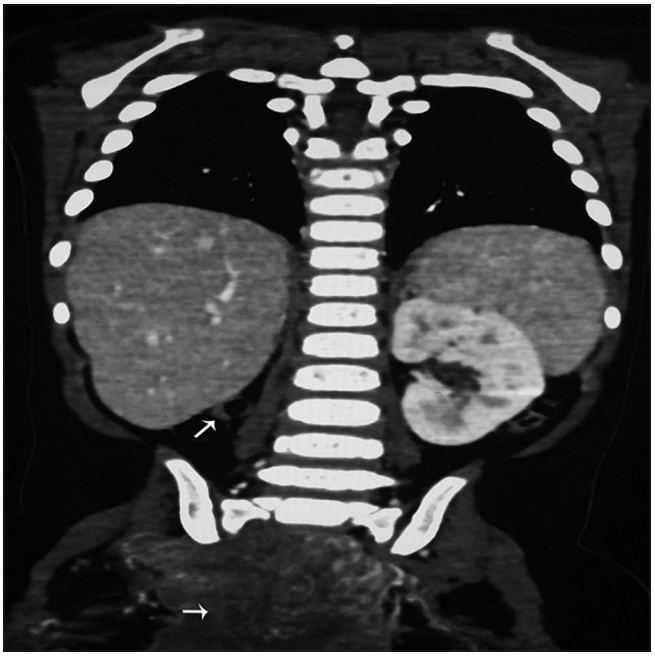

Figure 4: Coronal section of contrast-enhanced CT scan abdomen showing absence of right kidney in right renal bed and elsewhere in abdomen with compensatory hypertrophy of left kidney

Genital anomalies are more common in females accounting for 37–60%. The patient in this case had SSU. Septate uterus is the most common Mullerian duct anomaly, accounting for approximately 55% of the cases.[5] Its diagnosis is important because it is surgically correctable and has a strong association with repeated miscarriage. The most common diagnostic dilemma encountered is difficulty to differentiate between a septate and bicornuate uterus. The primary difference is the appearance of the uterine fundus; a septate uterus will have a normal convex external fundal contour. The septum itself originates from the midline of the uterine fundus and is a result of complete or partial failure of reabsorption of the uterovaginal septum. The septum may be partial or complete. A partial septum extends proximal to external cervical os while complete septum extends to the external cervical os, and may even extend into the vagina in approximately one-fourth of patients.[6] Although it is the most common uterine anomaly, it is notcommonly associated with URA. To our knowledge, no association of URA and SSU has been reported till date. Only one case of unilateral absent kidney has been reported with complete septate uterus along with obstructed hemivagina and unilateral agenesis of ovaries and fallopian tube by Mi Sun Kim *et al*.[7] Common genital anomalies associated with URA include uterine hypoplasia or agenesis, uni or bicornuate uterus, absent or aplastic vagina, absent fallopian tube and abnormal external genitalia. Renal anomalies are described in 40% of the cases with Mullerian aplasia and with unicornuate uterus.[8] Rolen *et al*.[9] found ipsilateral renal agenesis in 67% of patients with unicornuate uterus and pelvic kidney in 13%. Li *et al*.[10] reported renal agenesis in 30% of women with Mullerian duct anomalies and 80% of women with uterus didelphys. Ultrasound in combination with magnetic resonance imaging (MRI) plays an important role in diagnosing different abnormalities of female genital tract, especially uterine anomalies. To distinguish between septate/subseptate and bicornuate uterus MRI of the pelvis is the preferred modality. However, in this case, SSU was diagnosed on ultrasound and CT scan, MRI was not performed.

The patient in this report also had SCT. SCT is the most common germ cell tumor presented in newborns, occurring in 1 in 40000 live births. Females are more commonly affected than males with a ratio of 4:1.[2] Most of the SCTs are cystic and benign and only 1–2% are malignant.[11]

SCT are generally thought to develop from pleuripotential embryonic cells that has differentiated and matured. These primordial germ cells that deviated from their migratory pathway from the yolk sac to the gonad during early embryogenesis typically rest anterior to the future coccyx at Hensen's node. Growth of these primitive pleuripotential cells escapes the control of embryonic inductors and organizers, resulting in development of teratoma.[12]

SCT is associated with anorectal, vertebral and urogenital anomalies. Vogl and Riel[13] reported a case of Currarino's triad in which there is a combination of anorectal malformation and sacral dysplasia with a pre-sacral mass. In a study conducted by Lahdenne *et al*.[14] benign SCT was associated with vertebral abnormalities in 80% of the cases. The association of urogenital anomalies and SCT has not been widely reported. Shalaby *et al*.[15] reported a 12% incidence of urogenital anomalies, which were not apparent at the initial surgery and diagnosed later in life. Sahinoglu *et al*.[16] reported a case of urogenital sinus which was prenatally diagnosed in a fetus of type 4 SCT.

The treatment of SCT depends upon benign or malignant nature of the lesion and include removal of entire tumor and the coccyx bone. The chance of tumor recurrence is low with the removal of coccyx bone. Usually surgical removal of entire tumor is sufficient for benign tumors. However, individuals with malignant tumors receive additional treatment with chemo and radiation therapy.[12]

Conclusion

We report an unusual presentation of unilateral renal agenesis with SSU and SCT. Further exploration of this manifestation needs to be done in future to understand this unusual presentation and association.

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Conflicts of interest

There are no conflicts of interest.

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