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Case report

SUPERNUMERARY KIDNEY ASSOCIATED WITH OTHER GENITOURINARY AND CARDIOVASCULAR ANOMALIES IN A DYSMORPHIC INFANT: A RARE CASE REPORT

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ABSTRACT

Supernumerary kidney, also known as accessory kidney, is one of the rarest congenital anomalies of the genitourinary system without sufficient epidemiological studies. It is defined as having more than two kidneys with each kidney having its own vasculature, collecting system, and capsule. To date, there are few cases documented in literature worldwide. We hereby present the first reported supernumerary kidney case in a preterm 2-month-old infant with dysmorphic features associated with ectopic kidney and coarctation of the aorta. The coexistence of cardiovascular anomalies and genitourinary anomalies indicates a shared embryologic origin and genetic linkage, underscoring the importance of screening both systems when anomalies are identified in either. Newborn screening ultrasonographic examinations are crucial for detecting congenital anomalies, especially in infants with pertinent medical history or physical features, leading to better outcomes through timely diagnosis and intervention.

الملخص

الكلية الزائدة هي إحدى أكثر التشوهات الخلقية ندرة في الجهاز البولي التناسلي لذا لا تتوفر لها دراسات وبائية كافية. تعرف هذه الحالة بوجود أكثر من كليتين لكل منها أوعيتها الدموية, ونظام للقنوات الجامعة وغشاء خاص بها. حتى يومنا هذا, هنالك عدد محدود من الحالات الموثقة في البحث العلمي حول العالم. على ضوء ما تقدم, نضع بين أيديكم أول حالة كلية زائدة يتم الإبلاغ عنها في طفل خديج يبلغ شهرين من العمر بمعالم جسدية مشوهة مرتبطة بكلية منتبذة وتضيق بالشريان الأبهر. الوُجود المتصاحب للتشوهات في الجهاز الدوري الدموي والجهاز البولي التناسلي يشير إلى الأصل الجنيني والرابطة الجينية المشتركة بينهما. الفحص الأولي بالموجات فوق صوتية للمواليد مهم لاكتشاف مثل هذه التشوهات الخلقية, خصوصًا بين الأطفال ذوي التاريخ المرضي أو العلامات السريرية وثيقة الصلة, وقد يساهم ذلك في مخرجات صحية أفضل من خلال التشخيص والتدخل العلاجي في الوقت المناسب.

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1. INTRODUCTION

Congenital anomalies of the kidney and the urinary tract (CAKUT) manifest in various forms, with global prevalence ranging from 4.2 per 10,000 to 4 per 1,000 [1]. Despite CAKUT common occurrence, there is a paucity of literature specifically addressing supernumerary kidneys, also known as accessory kidneys. Supernumerary kidney is characterized by the presence of more than two kidneys and where each has its own vasculature, collecting system, and capsule. Like other genitourinary system anomalies, it is often associated with concurrent anomalies within the same system or other systems [2]. The scarcity of reported cases since Geisinger's review [3] of 40 cases in 1937 highlights the extreme rarity of this congenital anomaly. Due to the rarity of the cases reported in literature, no sufficient epidemiological data has been published. Rehder et al.; however, estimated the period prevalence of supernumerary kidney to be 1:26750 on CT imaging [4]. Most of the reported cases were found incidentally on imaging as supernumerary kidney is asymptomatic in two thirds (66%) of patients [4]. As the anomaly was found incidentally in most of the cases, it was not reported in the available literature until later in life when an imaging study was performed. In addition, there is a high potential for initial imaging misdiagnosis as supernumerary kidney is rarely encountered in clinical practice and deceivably could be interpreted as a more common pathology more likely by less experienced radiologists [4]. Given the aforementioned factors, no supernumerary kidney case was ever reported as early as in our patient. Not to mention, the associated dysmorphic features and cardiovascular anomalies were found in our patient.

2. CASE PRESENTATION

A 2-month-old healthy preterm boy underwent a screening newborn sonographic examination. He had unremarkable physical examination except for the facial dysmorphic features. Serum biochemical profile, routine blood workup and renal functional tests were within normal limits. Abdominal ultrasonography showed right ectopic pelvic kidney, left supernumerary kidney, and left native kidney where each had its own blood supply on color doppler (Figure 1).

Computed Tomography (CT) urography with contrast administration showed separated left two kidneys with normal size and a single right ectopic pelvic kidney (Figure 2).

The right ectopic kidney measured 3.5 cm in craniocaudal and 1.7 cm in mediolateral dimensions. Cranially located left kidney measured 3.2 cm in craniocaudal and 2 cm in mediolateral dimensions and had normal arterial supply and venous drainage. Caudally located supernumerary kidney on the left side was measuring 3.4 cm in craniocaudal and 2 cm in mediolateral dimensions.

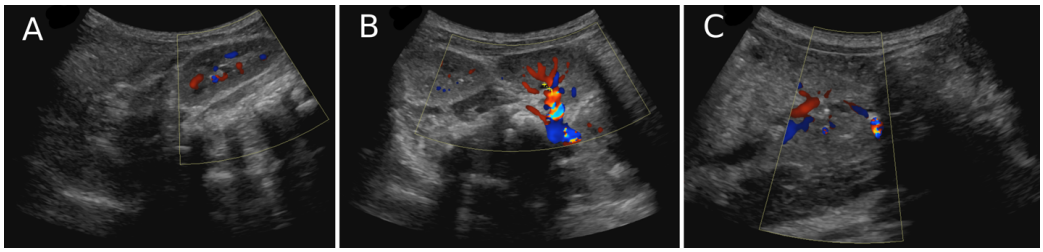

Figure 1 Abdominal doppler ultrasound images showing vasculature of all kidneys. (A) left native kidney, (B) left supernumerary kidney and (C) right ectopic kidney.

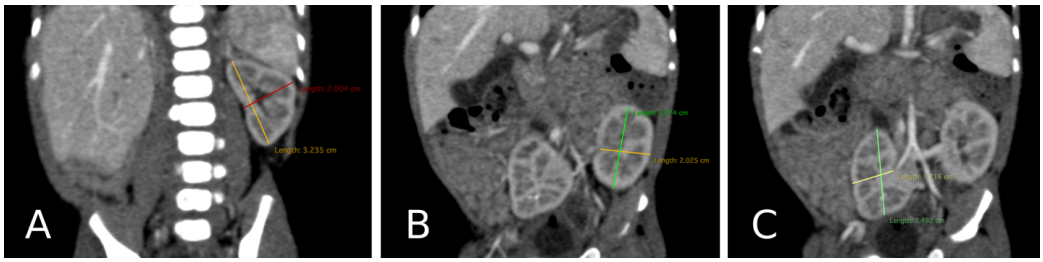

Figure 2 Coronal reconstructed computed tomography (CT) arterial phase images demonstrating three kidneys of normal size and shape. (A) left native kidney, (B) left supernumerary kidney and (C) right ectopic kidney.

The supernumerary kidney had a separate renal artery arising from the distal abdominal aorta (Figure 3A). It had two separated draining veins. The vein arising from upper pole unified with venous drainage of the left native kidney (Figure 3B). The vein arising from the lower pole unified with the thick venous trunk of the right ectopic pelvic kidney. Inferior Vena Cava (IVC) received as tributary only one thick venous trunk from the right side (Figure 3C). All kidneys showed normal homogenous nephogram enhancement and all renal veins were well opacified. The pelvicalyceal systems of all kidneys were not dilated and normally sited.

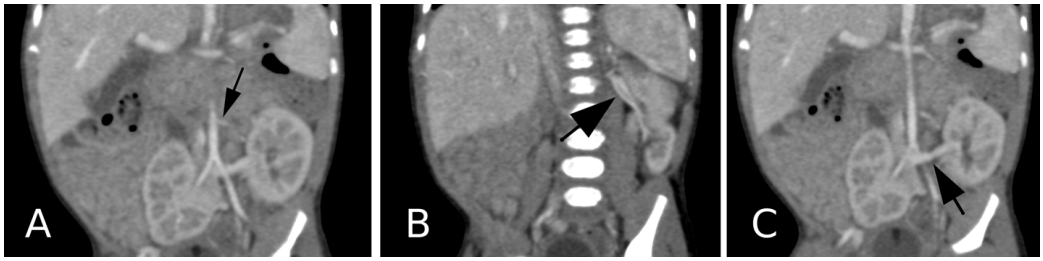

Figure 3 Coronal reconstructed computed tomography (CT) with contrast images of the left supernumerary kidney's vasculature. (A) showing renal artery (black arrow) emerging of the distal abdominal aorta, (B) showing unified upper pole venous drainage (black arrow) with the left native kidney's vein and (C) showing unified lower pole venous drainage (black arrow) with the right ectopic kidney's venous trunk.

On the contrast-enhanced CT study, an incidental finding of post-ductal aortic arch coarctation was documented (Figure 4).

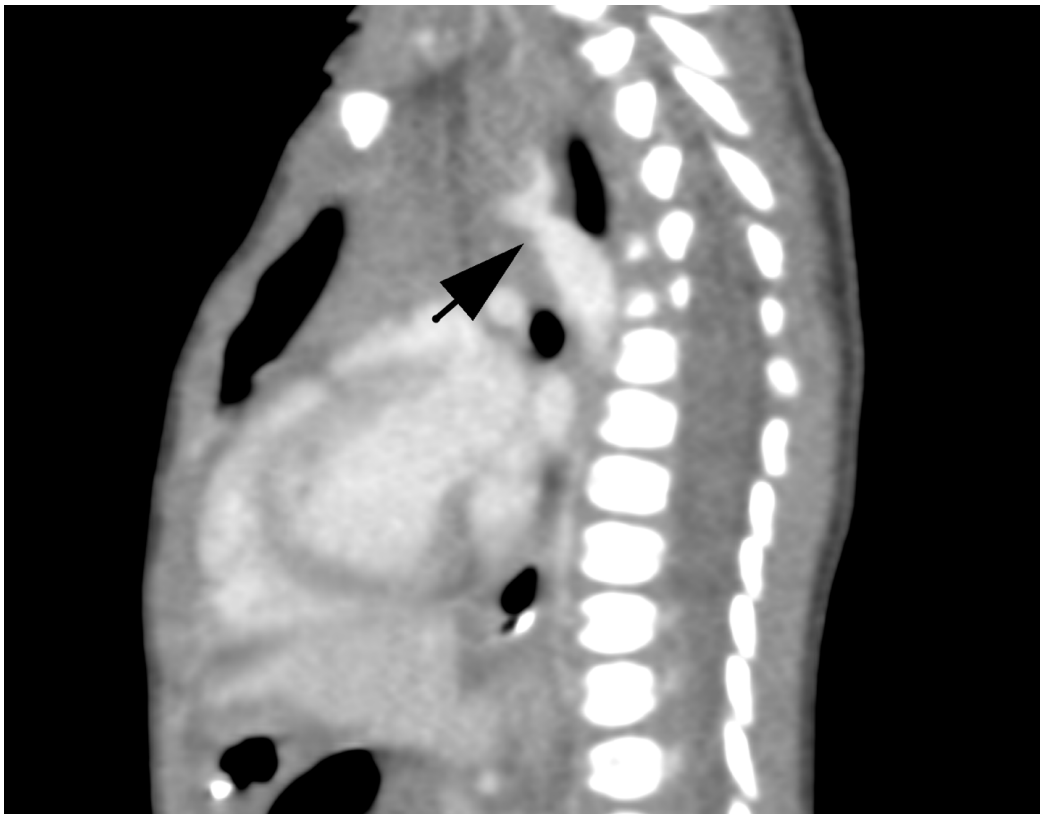

Figure 4 Sagittal reconstructed computed tomography (CT) arterial phase image demonstrating post-ductal aortic narrowing (black arrow).

3. DISCUSSION

A thorough understanding of the embryogenesis could contribute to a better comprehension of supernumerary kidneys and other associated anomalies [2]. Embryologically, nephrogenesis begins when the nephrogenic cord gives rise to the metanephric blastema making the kidney parenchyma, and the mesonephric duct gives rise to the ureteric bud making the renal collecting and drainage system. Although controversial, it is thought that supernumerary kidney could be a due to a division of either of these embryological origins giving rise to an accessory kidney [2].

In the past, most of the supernumerary kidney cases were diagnosed during autopsy or incidentally in surgery. Nowadays, increased awareness of the anomaly among radiologists, coupled with the utilization of advanced medical imaging modalities, has likely led

to a higher frequency of supernumerary kidney diagnoses. The establishment and compliance with newborn screening guidelines would potentially facilitate the early detection of such anomalies. While ultrasound (US) is a valuable screening tool for the renal system, its reliability in diagnosing supernumerary kidneys appears to be limited [4]. CT and MRI urography are the modalities of choice to more reliably assess both structural and functional features of the renal system [2, 4].

A supernumerary kidney must be differentiated from the more commonly look-like reported genitourinary anomaly, the duplex kidney [3]. Thus, a proper identification of the number, size, position, vasculature, and excretory system of the kidneys should be reported for the sake of documentation and, in case, an intervention is warranted.

While majority of the patients are symptomless, some may present with fever, flank pain, and urinary symptoms [4]. Although supernumerary kidney has no specific clinical implication in its own, the diagnostic misinterpretation of such anomaly warranted unnecessary interventions in some patients that led to increased morbidity [4]. Moreover, supernumerary kidney could be associated with complications such infections, stone formation and hydronephrosis. It, also, was reported with more serious conditions such as tumors and cysts [4]. Likewise, ectopic kidney found in our patient does not have a clinical significance unless complicated by other pathologies. It was, especially, found to be associated with nephrolithiasis and hydronephrosis. Furthermore, it could be misdiagnosed as other entities due to its different anatomic placement [5].

Congenital non-syndromic malformations, in general, were found to be more common among premature infants with no difference between both genders. Cardiovascular and genitourinary systems were the most commonly affected in (35.5%) and (27.7%) of infants respectively [6]. The associated cardiovascular anomaly (i.e. coarctation of the aorta) in our case was only reported once, according to our review of the literature [7]. This association between both systems could be explained by the shared embryologic origin of both systems, namely the mesoderm. There is a significant association between low birth weight and advanced maternal age at delivery with the co-occurrence of both system anomalies [8]. Children with congenital genitourinary anomalies had a tenfold higher reported cardiovascular anomalies [9]. Additionally, thirty percent of those affected with a congenital heart disease had an associated congenital genitourinary anomaly [9, 10]. Furthermore, a research team has reported that 29% of the mutagens associated with cardiovascular anomalies are also implicated in genitourinary anomalies, despite the fact that the majority of the underlying genes responsible for these conditions have not been identified yet [11]. Based on that, a potential genetic factor could be addressed and investigated in further studies. In addition, those diagnosed with a congenital anomaly of either of these two systems are advised to have a routine assessment for anomalies in the other system [9].

As supernumerary kidneys are, most often, symptomless and do not have any clinical implications on their own, patients mostly do not require any therapeutic intervention unless complicated otherwise. In such cases, supernumerary kidneys need to be considered as a differential as they could mimic other pathologies due to their unusual anatomic placement and unfamiliarity in the common clinical practice [4]. Depending on the suffered complication and the renal functional assessment, an appropriate intervention should be offered.

4. CONCLUSION

A supernumerary kidney is an extremely rare congenital anomaly of the urinary tract that typically goes undiagnosed until later in life. Our case report presented the earliest ever reported case of supernumerary kidney associated with other anomalies at an early infancy. Preterm delivery was found as an associated predicating factor for congenital non-syndromic anomalies. Cardiovascular anomalies are highly associated with genitourinary anomalies possibly due to their shared embryologic origin and a potential genetic linkage for both anomalies. Hence, a screening for both systems is advised if anomaly was found in any. This case highlights the usefulness of newborn screening ultrasonographic examination in diagnosing possible congenital anomalies especially among infants who have suggestive history (e.g., prematurity) or physical examination findings (e.g., dysmorphic features). Such practice might improve the clinical outcomes of patients by diagnosing congenital conditions and, if needed, treating them in a timely manner.

CONFLICT OF INTEREST

There is no conflict of interest to declare

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N/A

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