Congenital Giant Hydronephrosis: A Rare Cause for Upper Abdominal Mass in the Newborn

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ABSTRACT

We report a rare case of antenatally detected unilateral pelvi–ureteric junction obstruction leading to congenital giant hydronephrosis presenting as upper abdominal mass at birth.

Key words:
Antenatal ultrasonography, congenital giant hydronephrosis, newborn, upper abdominal mass

INTRODUCTION

Widespread use of routine antenatal ultrasonography (USG) has led to progressive increase in the detection of fetal anomalies. Antenatally detected urinary tract anomalies form a significant percentage of fetal anomalies and have an incidence of 2-9 per 1000 live births. Fetal hydronephrosis (physiological as well as pathological) constitutes 50-87% of the urinary tract anomalies detected antenatally.[1] Pelvi–ureteric junction obstruction (PUJO) remains the most common and important cause for pathological hydronephrosis during fetal life. Almost all cases of unilateral hydronephrosis detected antenatally are clinically silent at birth. Very rarely, gross hydronephrosis may be apparent at birth by virtue of its mass effect.[2] We report a rare case of antenatally detected PUJ obstruction with grade III Society of Fetal Urology (SFU) hydronephrosis[3] presenting as upper abdominal mass lesion at birth.

CASE REPORT

A 20-year-old primigravida with no known co-morbidity was referred to the radiology department for a routine antenatal USG at 20 weeks 3 days of gestation by her last menstrual period. USG revealed appropriate fetal growth corresponding to gestational age. The left kidney of the fetus demonstrated dilated pelvis with antero-posterior (AP) dimension measuring 8.6 mm [Figure 1]. Follow-up USG was performed 1 month later, which revealed the progression of hydronephrosis with AP dimension of pelvis measuring 19 mm along with dilatation of calyces (SFU grade III, Figure 2]. Amniotic fluid was normal in quantity. No other congenital anomaly was detected at this point. She had a spontaneous preterm (at 35 weeks) vaginal delivery of a low-birth weight (1960 g) male baby. At birth, the neonate had mild respiratory distress, which settled over the next few hours with conservative treatment and did not require any active resuscitation. The baby was slightly irritable with visible distension of the upper abdomen [Figure 3]. There was a palpable lump across the entire upper abdomen (epigastric, supra-umbilical, both hypochondrium and both flanks). The neonate underwent an urgent USG, which revealed hydronephrotic left kidney (SFU grade IV) with grossly distended renal pelvis (40 mm × 65 mm × 72 mm, AP × TR × CC, ~187 cc) extending across the midline to reach the right hypochondrium and flank [Figure 4]. The right kidney and urinary bladder were normal on USG. Hematological parameters including blood urea, serum creatinine and electrolytes were within normal limits. Initially, the neonate was managed conservatively as the vital parameters, including urine output, were normal. Micturating cysto-urethrogram was performed at a later date, and was found to be normal. The child underwent a radionucleide dynamic renogram at 2 months of age, which revealed dilated obstructed pelvis with compromised function of the left kidney. The left kidney demonstrated reduced perfusion with glomerular filtration rate of 74% and a split function of 37%. No excretion was noted on the left side during

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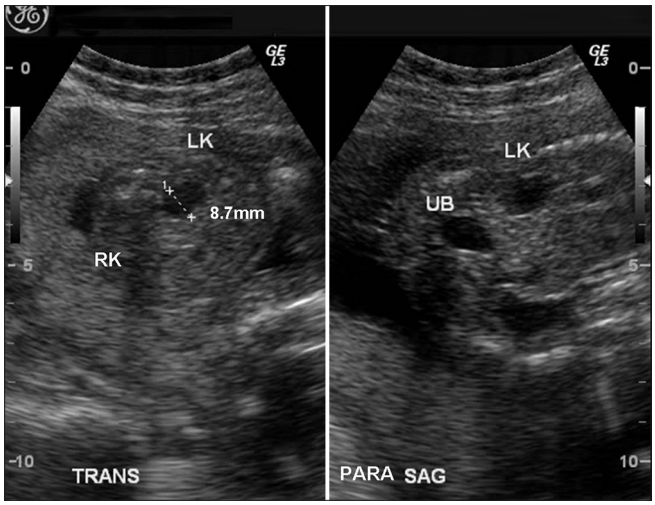

Figure 1: Antenatal ultrasonography of the fetal abdomen at 20.3 weeks of gestation: Transverse and parasagittal images depicting dilated left renal pelvis with AP dimension of 8.7 mm. (RK – Right kidney; LK – Left kidney; UB – Urinary bladder; TRANS – Transverse section; PARA SAG – Para sagittal section)

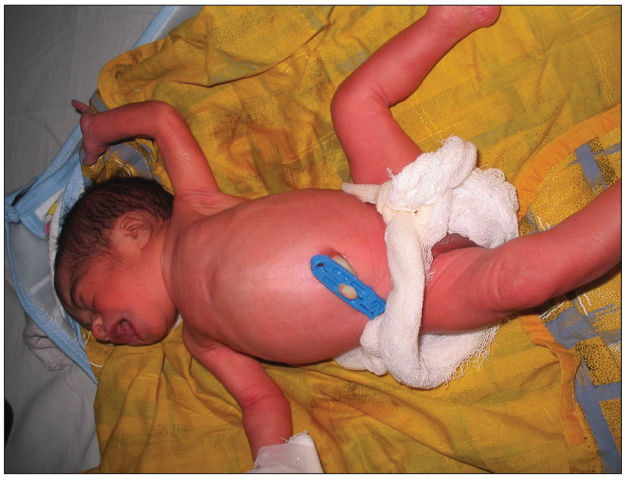

Figure 3: Day 1 neonate: Note uniform distension of the upper abdomen in a crying neonate

the dynamic renogram. Prolonged excretion was noted on the right side. A dimercapto succinic acid renal scan revealed enlarged left kidney with markedly reduced tracer concentration and persistent excretion through the right kidney [Figure 5]. The child was operated (pyeloplasty done) at the end of the 4th month. Intraoperatively, the findings of antenatal as well as post-natal USG were confirmed. Left renal pelvis was ballooned out and was reaching to the right side across the midline with thinned-out renal parenchyma. The child has been followed-up for 18 months and has been doing well. Follow-up USG revealed postoperative status with residual dilated PC system of the left kidney.

DISCUSSION

Over the last few decades, there has been constant increase in the discovery of fetal anomalies due to routine

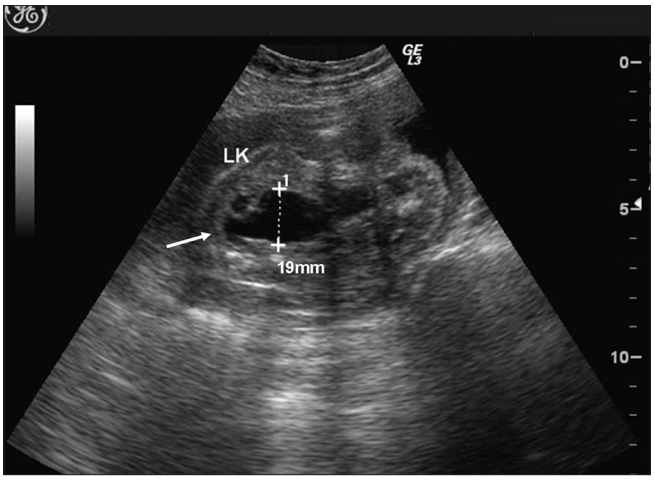

Figure 2: Follow-up ultrasonography approximatel 1 month later: Progressive increase in dilatation of the left renal pelvis with AP dimension of 19 mm. Note dilatation of calyces as well (white arrow) consistent with Society of Fetal Urology grade III hydronephrosis

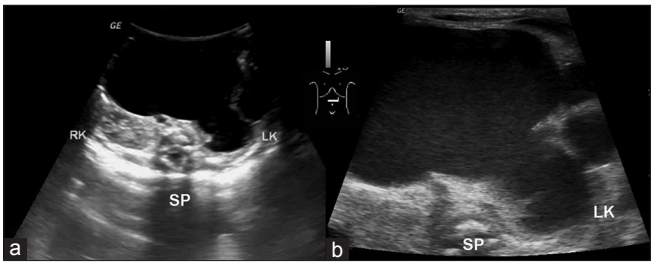

Figure 4: Day 1 ultrasonography: (a) Transverse section (5 MHz convex probe) of abdomen at the level of the renal hila. Note grossly hydronephrotic left kidney with dilated left renal pelvis reaching anterior to the right kidney across the spine (SP). The right kidney appears normal and (b) High-resolution image (10 MHz, linear probe) confirming the same findings

use of antenatal sonography across the globe. Varying degree of fullness of fetal renal pelvicalyceal system is a common finding during antenatal USG in the second and third trimesters. Over last two decades, a variety of scoring systems have been developed to grade fetal hydronephrosis and differentiate the physiological from pathological hydronephrosis.[3-7] The literature is abundant with long-term follow-up studies of antenatally detected fetal hydronephrosis. However, debate continues till today about conservative versus operative treatment and the timing of treatment of fetal hydronephrosis.[8-10] Although all of grade I and majority of grade II SFU hydronephrosis resolve spontaneously on conservative management and follow-up, a significant proportion of grade III and grade IV SFU hydronephrosis may need surgical intervention in the postnatal period.

Unilateral fetal hydronephrosis, even though pathological, is generally silent at birth and is not clinically apparent. In a study by Shimada *et al.*,[2] of 562 children treated for PUJO over a period of 14 years, only nine (1.6%) children required

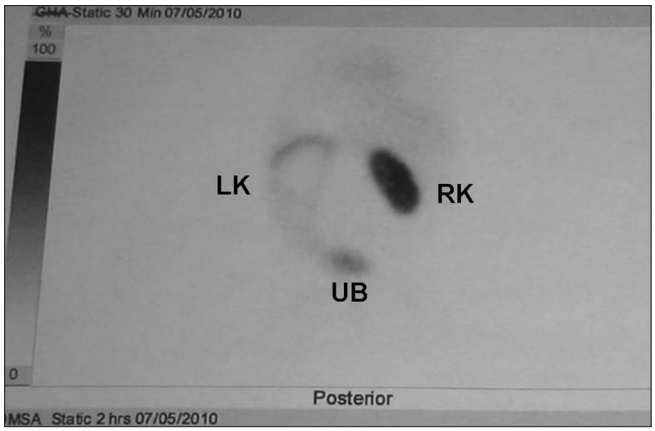

Figure 5: DMSA scan: Posterior view depicting enlarged left kidney with markedly reduced tracer concentration and persistent excretion through the right kidney at 2 hr static image

urgent urinary tract drainage in the neonatal period for renal dysfunction (5, 0.88%), giant hydronephrosis with mass effect (3, 0.53%) and severe urinary tract infection (1, 0.17%), respectively. It is noteworthy that of the eight fetuses detected to have unilateral hydronephrosis antenatally, contralateral kidney was normal only in three while the remaining five (62.5%) fetuses were diagnosed to have multicystic dysplastic kidney.

Congenital unilateral hydronephrosis presenting as ipsilateral palpable lump has been described in the literature. In the case described, hydronephrosis was so gross that the dilated left renal pelvis was seen to cross the midline and reach up to the right flank, which, to the best of our knowledge, has not been described in the literature. Uniform distension of the upper abdomen at birth with palpable lump in all quadrants of upper abdomen created a clinical diagnostic dilemma regarding etiology of the mass lesion. Despite prior knowledge from antenatal USG about left-sided PUJO with hydronephrosis, it was never expected to reach the right flank across the midline. We recommend that, although rare, the possibility of giant hydronephrosis should be included in the list of differential diagnosis of congenital upper abdominal mass lesion presenting at birth.

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