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Case report

Neonatal giant hydronephrosis – a rare case report

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Abstract
Introduction: Pelvi uretric junction obstruction (PUJO) is the most common cause of hydronephrosis in the neonatal period and is also the commonest cause of a palpable abdominal mass in a child. Giant hydronephrosis (GH) in a neonate is rare.
Observation: We are reporting a unique case of neonatal giant hydronephrosis which was managed successfully.
Conclusion: Establishing the correct diagnosis of GH is necessary to plan appropriate surgical intervention.

Introduction

With an overall incidence of 1:1500, PUJO is the most common cause of neonatal hydronephrosis. It predominantly affects male newborns. Exact incidence of GH is not known. Majority of reported cases occur in infants and children, and are congenital in origin. Surprisingly, definition of GH in neonates has not been defined yet [1,2].

Case report

A Primi gravida with an antenatally detected hydronephrosis was referred to us at 20 weeks of gestation. Antero posterior diameter of pelvis (APD) was 16 mm at 20 weeks, and follow up scans revealed progressive increase in the APD (Fig. 1A), 25 mm at 24 weeks and 60 mm by 34 weeks which progressed to 100 mm by 37 weeks. Baby was born by full term vaginal delivery, with a birth weight of 2.8 kg. Baby cried immediately after birth, was noticed to have tense abdominal distension with respiratory distress requiring oxygen. Post natal ultrasonography on day one of birth, revealed gross hydronephrosis with APD of 15 cm. Hence

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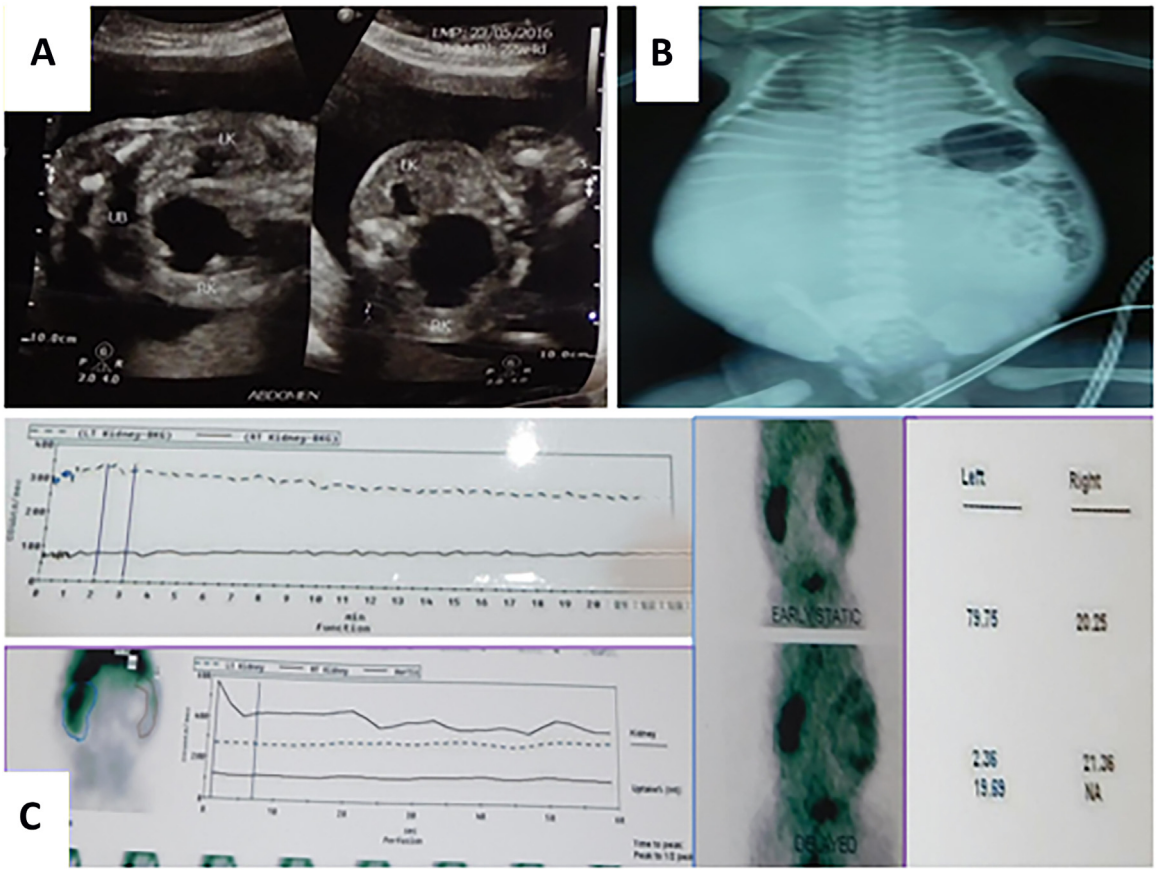

Figure 1 (A) Antenatal ultrasonography – showing features of hydronephrosis. (B) Postnatal erect abdomen X-ray showing. (C) Preopeartive radionucleide EC scan showing features of PUJO.

on day one of life Ethylene Cysteine (EC) radionuclide scan was done which revealed pelvi ureteric obstruction with differential function of 20% markedly impaired function as well as cortical tracer transit, due to PUJ obstruction (Fig. 1C). Echocardiography revealed moderate PDA and PFO with left to right shunt. Subsequently, follow-up 2D echocardiography revealed absolutely normal study.

In view of baby's increasing respiratory distress and huge abdominal distension (Fig. 1B), a surgical intervention was planned. Intra-operatively, we found hugely dilated renal pelvis containing more than 500 ml of clear urine occupying whole of the abdomen (Fig. 2D and E). A dismembered pyeloplasty (Anderson–Hynes) was performed through an extra peritoneal flank approach; the hydronephrotic pelvis was decompressed by wide bore 18 Gauze needle to minimize the spillage of urine, hence avoiding extensive dissection. 3french (FR) double J stent along with 10FR nephrostomy was placed (Fig. 2D&E). Anesthetic induction was done by inhalation sevoflourane, intravenous thiopentone and fentanyl. Internal diameter ID 3 mm endo tracheal tube was intubated. Thought out the surgical procedure muscle relaxation was maintained by intravenous atracurium and left to right shunt reversal factors especially hypoxia and dehydration were avoided by maintaining adequate oxygenation as well as hydration. Subsequently, intravenous neostigmine and glycopyrolleate were used for extubation and reversal. Baby recovered well post operatively. Nephrostomy was clamped after 48 h and removed on day 5. JJ stent was removed after 8 weeks. Follow up radionuclide scan at 6 months revealed renal function of 43% with good clearance (Fig. 2F). Baby is on follow up since one year with us.

Discussion

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 amongst which Pelvi–ureteric junction obstruction (PUJO) remains the most common. 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 [1].

Many definitions for GH have been reported in the literature, but none in neonates. Historically, only 50% of GH cases are properly diagnosed because of its non specific clinical presentation. There are no long-term follow-up and outcome data in the literature on GH available especially in neonates including its definition [2].

In 1939, Sterling first defined GH as draining more than 1 l fluid or fluid amounting to 1.6% of body weight in the collecting system. Later on, radiographic criteria for GH were defined by Crooks et al.

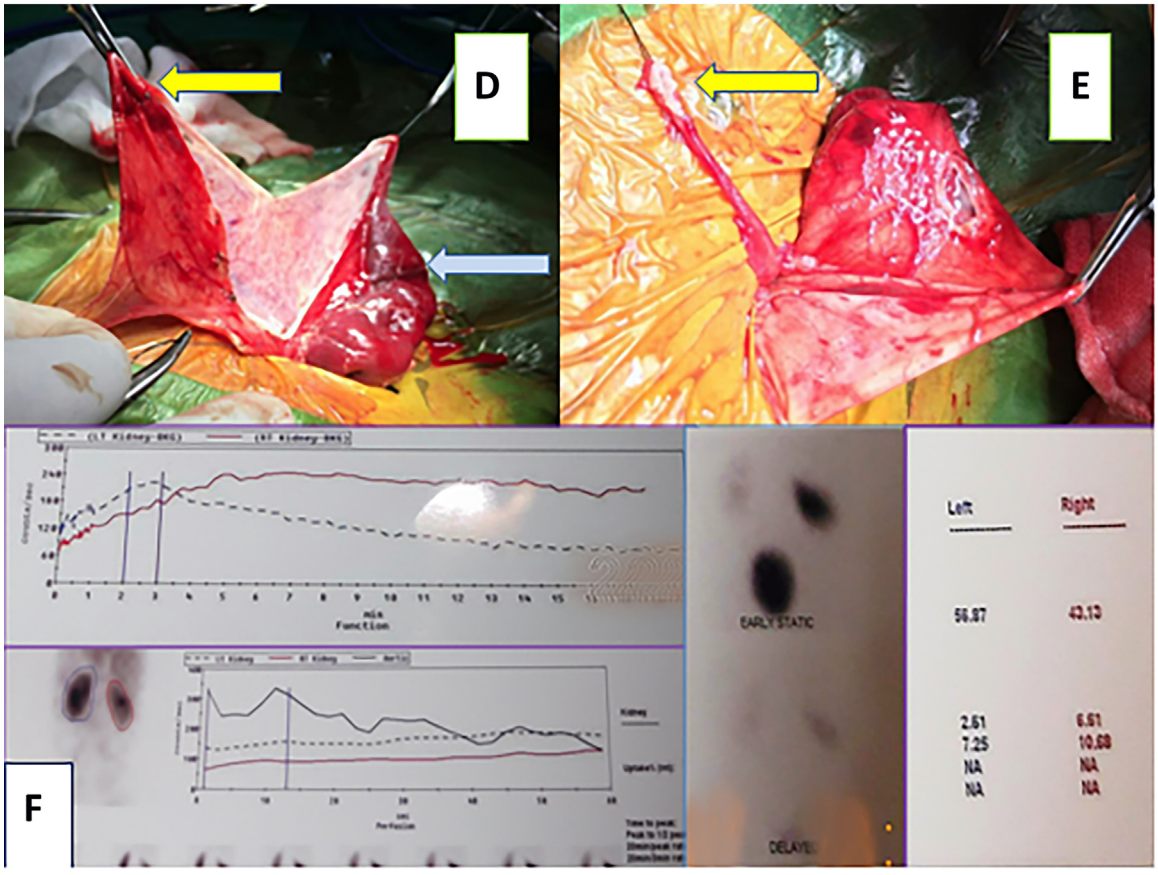

Figure 2 (D and E) Intraoperative picture showing dismebered PUJO. Yellow arrow pointing at PUJ. (F) Post operative radionucleied EC scan.

as the kidney occupying the hemi-abdomen which also meets or crosses the midline and has a height of about 5 vertebral bodies [3,4]. Yang et al. (1958) opined that the term GH should be used only when the contents of the sac equaled to average daily urine output for that age [2].

On literature search and review, we found various ways of management of neonatal PUJO; but very few reports on neonatal GH.

The first case of GH was published in 1746, and since then less than 500 cases have seen described worldwide to date, with most cases reported within the last 15 years [5].

As reported by various studies, if renal function (RF) is severely impaired by 5–20%, percutaneouss nephrostomy and subsequent pyeloplasty if it improves the drainage function or else nephrectomy. Pyeloplasty reserved for 20–40%renal function, where as >40% RF are managed expectantly with regular follow up [6].

Several published series have supported immediate pyeloplasty in infants with prenatally diagnosed hydronephrosis and confirmed postnatally as PUJ obstruction [7].

However, a puncture/drain procedure may also be performed in cases in which the condition of the patient does not allow other treatments or when hemodynamic changes can occur following a sudden abdominal decompression [8].

Hoffman preferred nephrectomy in kidneys affected by GH as there was no improvement in function, in addition to higher gastrointestinal disturbances and increased susceptibility to trauma caused by the retained hugely hydronephrotic kidney [8].

Rapid and safe decompression of the obstructed system by ultrasound guided Percutaneous nephrostomy performed over JJ stent has also been reported previously in the literature by Ahmad et al. [9].

We had a neonate with antenatally diagnosed hydronephrosis at 20 weeks, which progressively increased to term with APD of 100 mm, and had huge abdominal distention compromising its breathing. Hence immediate pyeloplasty was done at day 2 of life without any events intra operatively and post operatively. Since immediate postal day 2 pyeloplasty has not been described in the literature so far; we are reporting it in view of its uniqueness and its surgical challenge.

Conclusion

Neonatal giant hydronephrosis is rare. Establishing the prompt antenatal diagnosis of GH is necessary to plan appropriate postnatal surgical interventions. Immediate postnatal primary pyeloplasty is surgically challenging; but do yield good results. This procedure avoids morbidity of a staged procedure, as it was witnessed in our case, though gratifying long term results are awaited.

Conflict of interest

Authors declare none conflict of interest.

Authors' contribution

JSA: concept, collecting the data, collecting the literature, review of literature, writing the article, grammatical corrections of the article, preparing the pictures, uploading the article and pictures; MNB: operating on the patient, grammatically correcting the article; VJ: assisting in operating on the patient; VKK: anesthesiologist for the case.

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Consent from the Patient

A written consent was obtained by the parents.

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