Case Report

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Early pyeloplasty and excision of a multicystic kidney in a neonate with a complicated nephrostomy: A case report

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Introduction and Importance: The association of multicystic renal dysplasia with ureteropelvic junction obstruction is noteworthy in newborn. However, the possibility of conservative management is still a primary requirement, unless there are complications that suggest surgery. The authors are studying a case of a newborn in which an incorrect nephrostomy led to complications that required emergency surgery.

Case Presentation: A newborn girl with left-side ureteropelvic junction obstruction and an enlarged and multicystic right kidney was operated on at an early age by unexpert hands, leading to complications. Was monitored daily, and an emergency procedure was done. Following up shows the success of the emergency operation.

Clinical Discussion: The age and the precise timing of intervention are controversial. In this case, several postnatal diagnostic tests were made due to the severity of the hydronephrosis in the antenatal period, which resulted in the implementation of percutaneous nephrostomy.

Conclusion: Authors suggest that it is better to not operate as long as the patient's condition is stable

Keywords multicystic kidney, neonate, nephrostomy, pyeloplasty, ureteropelvic junction obstruction

HIGHLIGHTS

- Multicystic renal dysplasia is one of the most common renal malformations that affects neonates.
- Surgical interventions should be delayed as much as possible to minimize complications.
- It is possible and under emergency conditions to perform a complete surgery in which the obstruction is corrected.

Introduction

Multicystic renal dysplasia (MCRD) is one of the most common urinary tract abnormalities in children and the second most common cause of kidney masses after hydronephrosis, which is often unilateral[1,2].

MCRD is mostly idiopathic, but some research suggests that it may result from a significant defect in urine flow in the early stage of renal development. Where the renal parenchyma is replaced by nonfunctional renal tissue[1,2].

On the other hand, ureteropelvic junction obstruction (UPJO) is recognized as the most common defect of the upper urinary tract in children, also, it is the second most common abnormality in the context of MCRD after contralateral vesicoureteral reflux[3,4].

The diagnosis is made for both cases through the ultrasound during pregnancy, and when the defect is noticed the fetus is followed up until birth[1,2].

As for management, previously, polycystic kidneys were managed by surgical excision. Currently, doctors recommend conservative treatment unless any complications or abnormalities require surgery[5,6].

Nonoperative follow-up is generally recommended for the postnatal management of UPJO, especially unilateral mild to moderate hydronephrosis. Whereas, in severe hydronephrosis patients, indications of the type and time of the early surgical intervention are debatable[7].

Extraperitoneal resection pyeloplasty is the preferred treatment for UPJO, especially when renal function is impaired, but when a severe obstruction is not present or the patient does not need definitive repair, a temporary percutaneous, tube or percutaneous pyeloplasty may be performed with an assessment of renal function pending. It will be fixed later, as using this technique depends on monitoring the renal cortex thickness and the diameter of the renal pelvis[5,6].

Anesthesia and surgery are safe for healthy newborns if performed by an experienced hand[8].

In our case, the child had MCRD combined with UPJO. A nephrostomy was performed to manage the case, which led to complications in the child and she needed emergency surgery in which a pelvic-ureteral junction was made with a polycystic nephrectomy at the same time, and the baby's condition improved after that.

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Methods

This case report has been reported in line with the SCARE criteria 2020[9].

Case report

History

A pregnant woman in the fifth month visited the obstetric clinic for a regular follow-up and an ultrasound was performed which showed the following: dilatation of the cysts and pelvis in the left kidney, cystic cortical formations of different diameters in the right kidney. Before the pregnancy, the mother did not take any kind of medication, while during pregnancy she was taking folic acid supplements. There were no complications in the family history, and a mild posttraumatic stress disorder was diagnosed, related to war events in the region. No complications were faced in the delivery, and 5 days after delivery, an ultrasound was performed showing an enlarged and multicystic right kidney, in addition to obstruction of the pelvic-ureteral junction. Laboratory tests showed an increase in creatinine to 3.16 mg/dl. The next day, a contrasted multislice was performed, which showed dilatation of the left emptying apparatus and a large number of cystic formations in the right kidney (Figs 1, 2).

At the age of 1 week, a left nephrostomy was conducted by nonexpert hands at another hospital, which is not recommended, thus, the case is complicated with leakage into the abdomen and the expansion of the abdomen.

Physical examination and other investigative modalities

A 9-day-aged newborn girl was referred to our hospital, and a dilated abdomen was noticed at the physical examination with a left nephrostomy. Lab tests showed a continuous increase of creatinine along with high urea in addition to high C-reactive protein and leukocyte values and a low sodium value. Ultrasound of the urinary system was performed the next day, and showed the right kidney with many large cysts with no distinguishing of the renal cortex, whereas, the left kidney had an enlargement of the excretory system and the pelvis with the presence of a hematoma in the lower pole.

Detailed treatment plan

The decision was made to do an emergency operation, as a result of the elevated creatinine levels with abnormal laboratory test values and the deterioration of the child's condition, since the right kidney is unfunctional.

On the 11th day after delivery, the intervention was made through a left lumbar intraperitoneal incision, by a team of pediatric surgeons and urologists. A surgical nephrostomy was performed with a pyeloplasty on the left side, in addition to the excision of the right polycystic kidney through the same incision, while the right ureter diameter was 1.5 cm.

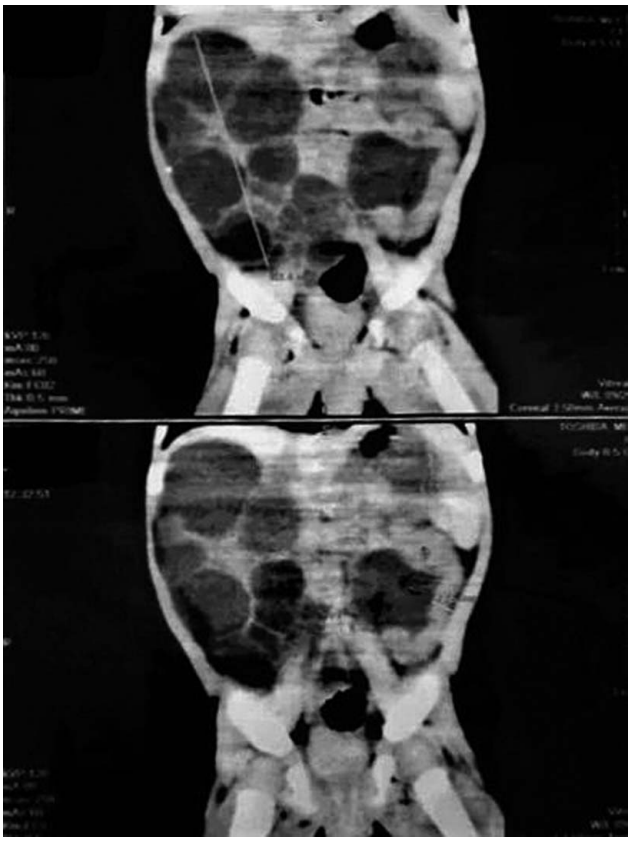

Figure 1. Coronal section computed tomography images showing the large multicystic kidney.

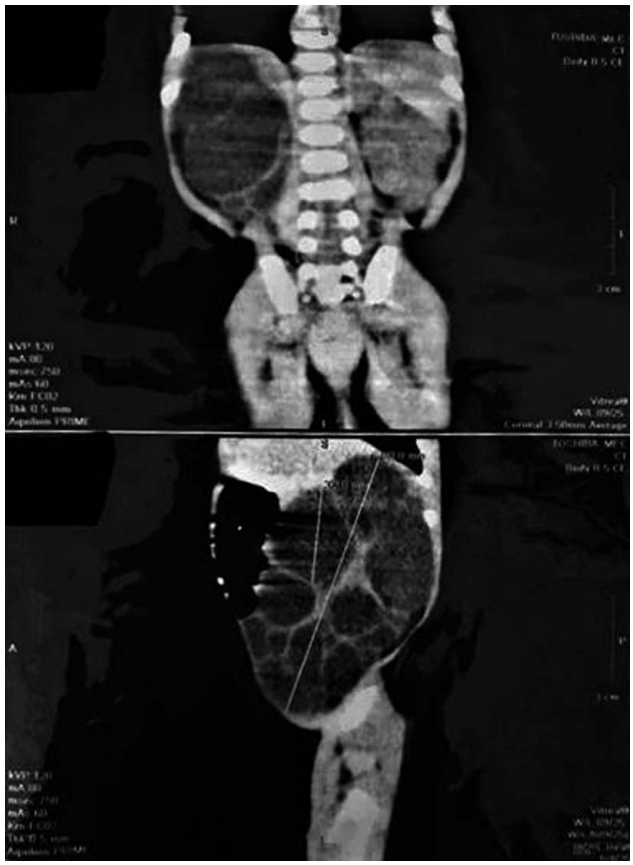

Figure 2. Coronal and sagittal sections computed tomography images showing the large multicystic kidney.

Follow-up and outcome

Postoperative management essentially depends on the follow-up, an ultrasound of the urinary system showed no hematoma in the place of the excised right kidney. The leakage of the urinary into the abdominal cavity has stopped.

A contrasted radiography kidney image was made through nephrostomy, which showed no complications (Fig. 3).

Laboratory tests also showed that creatinine values decreased from 4.4 to 1.1 mg/dl, and the girl's condition improved within 2 weeks after the emergency operation (Table 1).

Table 1

Laboratory test values changes with time.

Age (days)	Urea	Creatinine	Sodium (Na)	Potassium (K)
5	77	3.16	-	-
9	113	3.2	120	4.37
11	115	4.4	107	4.44
14	94	3.1	125	3.0
16	83	2.0	136	5.4
17	90	1.85	130	5.1
20	64	1.1	138	5.2
Normal ranges	10–40	0.3–0.7	134–148	3.7–5.9
Units	mg/dl	mg/dl	mmol/l	mmol/l

Discussion

MCRD is one of the most common renal malformations that affect neonates, it is most commonly present with a restriction in the other kidney ureteropelvic junction[1,2], however, the precise timing of intervention is debatable.

The postnatal evaluation in infants with antenatal hydronephrosis is not agreed upon, as it is not always a result of an important obstruction[10]. In our case, several postnatal diagnostic tests were made due to the severity of the hydronephrosis in the antenatal period, which resulted in the implementation of percutaneous nephrostomy.

Nephrostomy can have many different complications, such as urinary leak into the abdomen, self-limited postprocedural bleeding into the pelvicalyceal system, chronic microscopic hematuria, urinary tract infection, dislocation of the PCN, cellulitis at the skin entry, urinary leak, and mechanical damage, with the latter three being the most common as described by Ključevšek *et al*[11]. Intra-abdominal urinary leak and an intrarenal hematoma were noticed in our case, in addition to the failure of renal function improvement, as the nephrostomy was performed by an inexperienced physician.

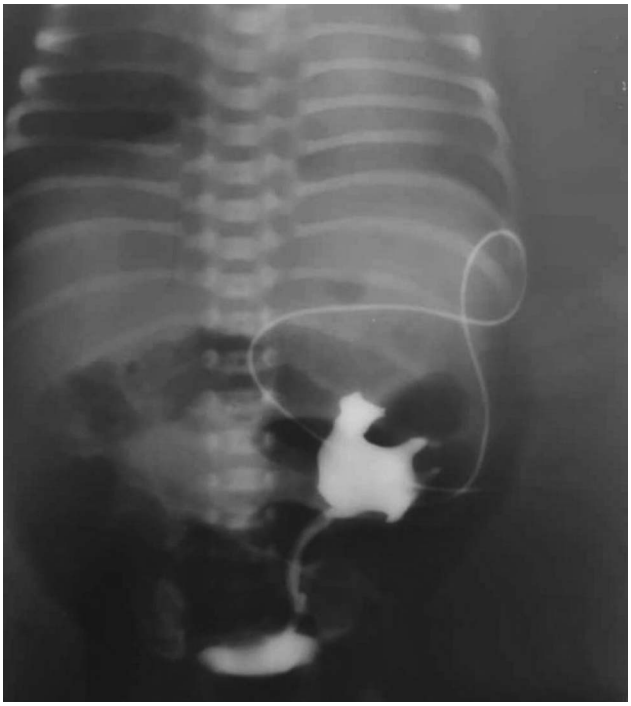

Figure 3. Anterior contrast media administration through the surgical nephrostomy (follow-up).

The neonatal period is sensitive; therefore, surgical interventions should be delayed as much as possible to minimize complications. However, if the benefits weighed the risks, pyeloplasty could be performed before 6 weeks of age. According to the Society of Fetal Urology grading system, there was no recommended grade for intervention[7,12]. Our patient was grade III according to the Society of Fetal Urology grading system with rapid renal function deterioration, and due to the sensitivity of the situation of having a single functioning kidney, an emergent pyeloplasty was indicated to spare the remaining kidney function. Moreover, intraperitoneal urinary leakage leads to a surgical intervention via the peritoneal cavity. The decision to remove the right nonfunctional multicystic kidney was made due to its large size and the implementation of intraperitoneal surgery.

Conclusion

This case provides additional evidence that we should observe and not operate at the early age on UPJO patients as long as the patient's condition is stable, as in our case, the early intervention led to a life-threatening situation. Also, it is possible and under emergency conditions to perform a complete surgery in which the obstruction is corrected and the polycystic kidney removed.

Ethical approval

Ethical approval was not required.

Patient consent

Written informed consent was obtained from the patient's parent for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

H.R., O.H., S.A., and F.K. drafted the first manuscript. A.K.D. and H.R. revised the final manuscript. R.A. performed the

procedure. R.A., A.K.D., and H.R. collect the relevant data from the patient.

Conflict of interest disclosure

The authors declare no conflict of interest.

Research registration unique identifying number (UIN)

This is not an original research project involving human participantsin an interventional or observational study but a case report. This registration was not required.

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