Giant hydronephrotic kidney in adolescence

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KEY WORDS

kidney ◗ hydronephrosis ◗ nephrectomy

ABSTRACT

Giant hydronephrotic kidney is a rare form of obstructive uropathy in adolescents. The authors report a 14 year old girl with large distension of the collecting system of the right kidney secondary to obstruction of the ureteropelvic junction. The cystic mass occupied the right retroperitoneal space and crossed the midline. The course before hospitalization was asymptomatic. There was only one symptom of a sense of stomach satiety. Nephrectomy was performed without complication.

INTRODUCTION

Giant hydronephrotic kidney was defined by Stirling in 1939 as a dilatation of the renal pelvis by more than one liter of fluid contained in collecting renal system [1]. This definition was widened in radiological examination to crossing the midline in transverse dimension and longitudinal section more than 5 vertebrae. Ureteropelvic junction (UPJ) obstruction is the most common etiology. Other rare causes are vesicoureteral reflux, megaureter, and posterior urethral valves.

CASE REPORT

A 14 year old girl was referred by her GP (general practitioner) to the gynecology and obstetrics department due to a large abdominal cystic mass, spanning from the suprapubic area to the area below the xiphoid process. The initial suspicion was a large ovarian cyst. On physical examination, ultrasonography, and computed tomography (CT), the ovarian pathology was excluded. She was sent to our Pediatric Surgery and Urology Department with the diagnosis of a multicystic dysplastic kidney.

On physical examination, a large painless cystic mass was palpable. It occupied the right hemi-abdomen and crossed over to the left. In the case history she never had flank pain or recurrent urinary infections. Blood pressure and hematology tests were normal. There was only one symptom, a sense of stomach satiety.

Computed tomography (Fig. 1) showed a fluid density, huge cystic mass without pathological vascularization. It was divided by a few thin septa. It was 22 cm in transverse, 10 cm in sagittal, and 30 cm in longitudinal dimensions. It occupied the entire right retroperitoneal space. The peritoneum was compressed and moved to the left but there was no intestinal obstruction. The liver capsule was modeled. Left kidney structure and function was normal but the right kidney structure and contrast excretion were not visible. The scans were done to confirm a nonfunctioning right kidney.

On the basis of the CT, the diagnosis of a giant hydronephrotic kidney was suggested, but we considered other possible causes like cystic neoplasm.

The girl was qualified to open nephrectomy. The lower medial laparotomy approach was chosen. After exposing the large cystic mass, a needle was inserted and the cyst was drained. Six and a half liters of urine were collected and the cyst was decompressed creating a large operating space. This maneuver allowed dissecting the cysts with great ease from surroundings tissues. The renal hilum had normal vessels. The ureter was significantly stenotic at the UPJ, which indicated a primary obstruction. The pelvis was small but calices presented massive dilatation (Fig. 3). Most of the collecting system was situated intrarenally and covered by marginally thin renal parenchyma (Fig. 4). During surgery, blood loss was minimal. Broad-spectrum antibiotics were given for 3 days postoperatively. Recovery was uneventful and the patient was discharge after the 4th day.

DISCUSSION

Giant hydronephrosis is rare form of obstructive uropathy, especially in children and adolescents. Comparatively large sizes of kidneys are encountered in newborns and infants where the kidney is larger than other parenchymatous organs. The distended kidney is palpable through the poorly developed abdominal wall. In older children symptoms of recurrent urinary infections, intermittent flank pains with vomiting and nausea, nephrolithiasis, hematuria, and blood hypertension dominate. In our case, the course was asymptomatic revealing a slow and chronic disease process.

We were surprised that the only symptom of hydronephrosis in this 14 year girl was abdominal distension. We encountered similar cases in our literature review. E. Yilmaz et S. Guney presented a 12 year old girl, who was admitted to the hospital due to progressive abdominal distension [2]. During surgery, 13.5 liters of urine were drained. However she had recurrent abdominal pain 4 months before admission to the hospital. A. K. Hemal et al. reported a group of 17 patients with giant hydronephrosis who underwent laparoscopic nephrectomy due to an abdominal mass [3].

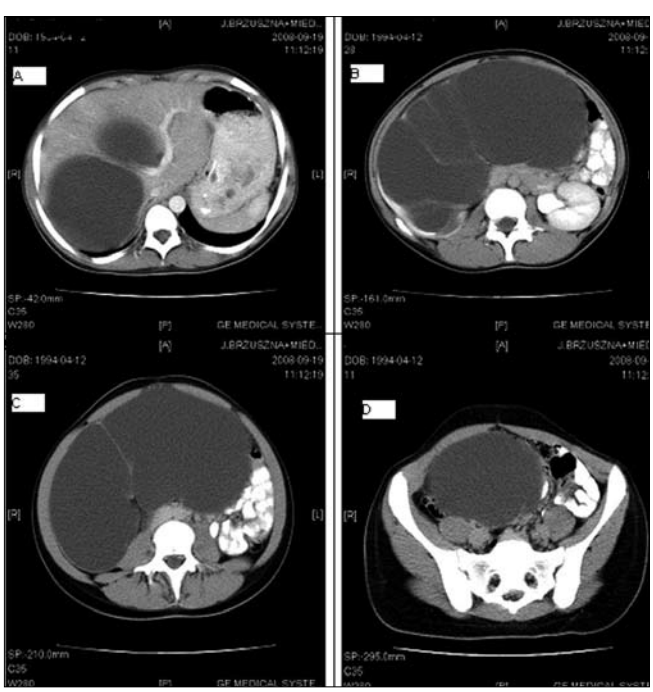

Fig. 1. CT images with intravenous contrast. Huge non-contrasted fluid-filled mass pressing the liver (A), occupying the right retroperitoneal space, crossing the midline (B, C), and ending in the pelvic cavity (D).

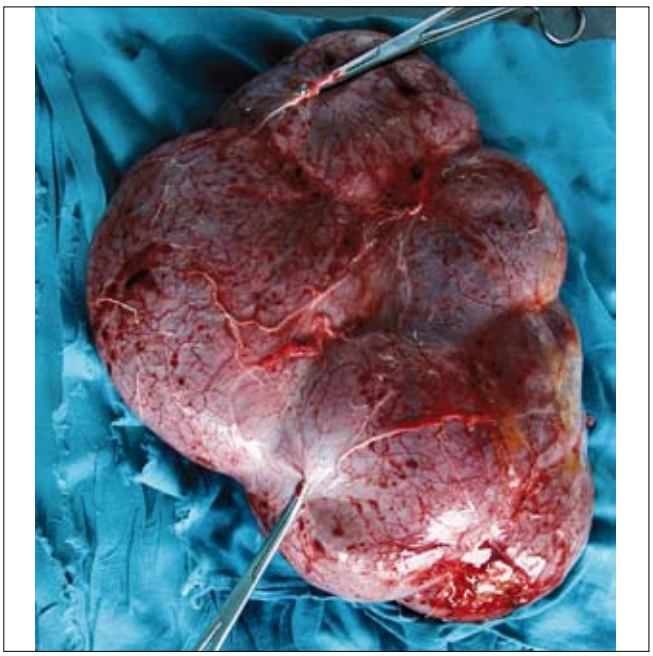

Fig. 2. Giant hydronephrotic kidney after extraction. The collecting system contains 6.5 liters of urine.

Huge distension of the collecting system poses the potential risk of major complications. Rupture of a giant hydronephrotic kidney after weak blunt trauma, like compresion by a seat belt, can easily be expected. Ipsilateral ileofemoral vein thromboses due to compression have been reported [4].

It makes us think about how it is possible to develop such an extent of pathology where symptoms are secondary to mass effect and are recognized late. The initial diagnosis of ovarian cyst is similar to Hoffman's historical report. In an era before the development of modern diagnostic imaging tools, giant hydronephrosis was misdiagnosed as an ovarian cyst, mesenteric or pancreatic cyst, ascites, or a renal tumor [5].

Most reported cases showed an dysfunctional kidney and were qualified for nephrectomy. Conservative treatment or salvage procedures had been reported in solitary or bilateral kidney disease. In cases, where the greater part of the distended collecting system is positioned intrarenally, typical pyeloplasty is not adequate and does not resolve the problem of urine retention. The volume of the collecting system is not reduced sufficiently because the biggest part is located in the calyces. Pyeloplasty in conjunction with nephroplication and nephropexy is a recommended solution for this problem. In this procedure, the floppy kidney is folded onto itself, bringing the lower pole to the upper pole in U-shaped configuration. It decreases the size of the calyces and nephropexy ensures gravity drainage. An alternative treatment is lower pole ureterocalycostomy, but it emphasizes a secondary obstruction after the procedure [6].

Laparoscopic nephrectomy for giant hydronephrosis has been reported in few articles. In Hemal's group of patients who underwent laparoscopic nephrectomy, the greatest volume of fluid aspirated was 5 liters in adult men, while the others had from 1.2 to 2.6 liters [3]. In our case, open laparotomy was chosen but, based on this experience, the laparoscopic approach was also possible. After suction of urine, the operating space was more comfortable and we achieved easier visualization and preparation circumstances. Additionally, it was easier to divide the kidney from the surrounding tissues. In the future, in similar cases, the laparoscopic nephrectomy will be implemented.

Differential diagnosis should consider teratoma, cystic neuroblastoma, partially differentiated cystic nephroblastoma, multicystic dysplastic kidney, or other cystic mass in the retroperitoneal space. However, hydronephrosis due to UPJ obstruction is possible in cases without solid areas in CT or MRI (magnetic resonance imaging) examinations.

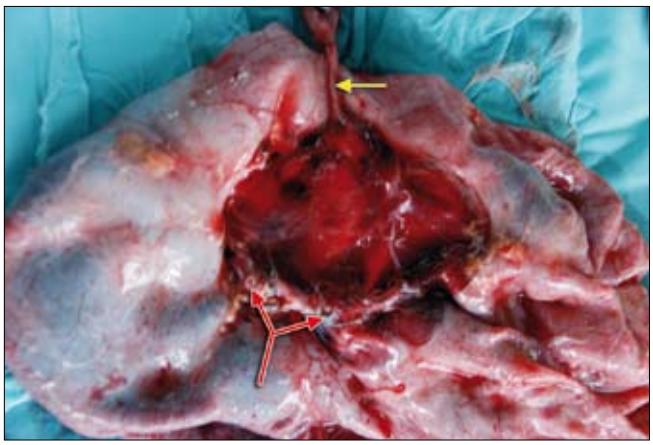

Fig. 3. Renal pelvis has no extrarenal dimensions. Collecting system is situated intrarenally. Indicated ureter (yellow arrow), renal vessels (red arrow).

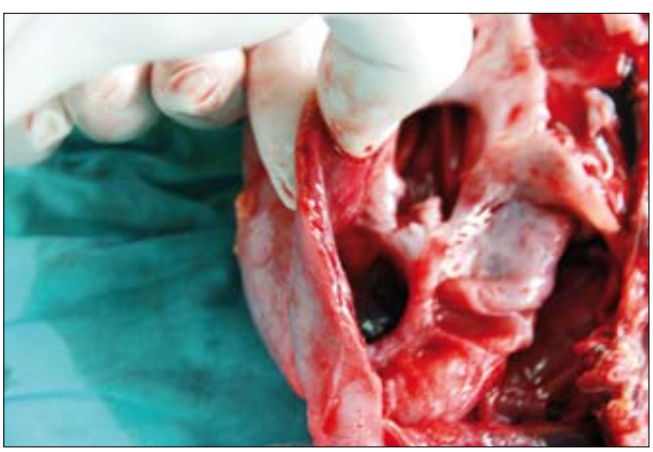

Fig. 4. Floppy kidney section showed thinned renal parenchyma and distorted calyces.

CONCLUSION

With improved diagnostic methods for hydronephrosis, we can even recognize very neglected cases.

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