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GIANT HYDRONEPHROSIS IN A PELVIC KIDNEY

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ABSTRACT

Pelvic ectopic kidney associated with giant hydronephrosis in children is rare. Clinically the pelvic ectopic kidney is usually asymptomatic or revealed by urinary symptoms. The diagnosis is easily made on an ultrasound scan. We report an atypical case of pelvic ectopic kidney with giant hydronephrosis that was discovered as a pelvic mass and for which the abdominal-pelvic ultrasound revealed a pelvic pseudotumor. The diagnosis was confirmed only by the CT scan. The aim of this case report is to show that particular diagnostic approach and to assess the radiological characteristics of that rare anatomical variant.

KEYWORDS: pelvic ectopic kidney, giant hydronephrosis, pseudotumor, abdominopelvic ultrasound.

INTRODUCTION

The pelvic kidney is a rare renal ectopy with an incidence of 1 in 12,000 births.[1] Pelvic kidney is usually asymptomatic but often associated with urologic abnormalities. Association of pelvic kidney and giant hydronephrosis is exceptional in children and a few reports have been published in literature.[2] The diagnosis can be easily established by ultrasound. We report a case of giant hydronephrosis in a pelvic kidney associated simulating a tumor to determine its radiological characteristics.

CASE DESCRIPTION

A 10-year-old girl was hospitalized in the Paediatric Oncology department and referred to the Radiology department for exploration of a pelvic mass that had been growing for 11 months. She had been complaining of abdominal-pelvic heaviness for several months. In clinical

examination, the child's overall condition was altered. There were no external genitalia disorders. Palpation have revealed a painful and non-mobile pelvic mass. There was also swelling and oedema of the leg. In the past, we have noted a history of repeated urinary tract infections. The biological screening showed renal failure with a creatine at 189 umol/L; urea was 2.90 mmol/l. Ionogram was normal with a natraemia at 149 mmol/l, kalaemia at 3.7 mmol/l and chloraemia at 101 mmol/l. Urinalysis noted leukocyturia.

An abdominal-pelvic ultrasound was performed, showing a heterogeneous pseudomass measuring 125 x 115 mm in size (Figure 1a), there was cystic and a soft-tissue portion. The soft tissue was vascularized in color Doppler (Figure 1b). Its origin was difficult to determinate due to its large size. The right kidney was not seen in its normal position and the left kidney appeared normal; there was no obstruction of urinary tract. There were no secondary lesions.

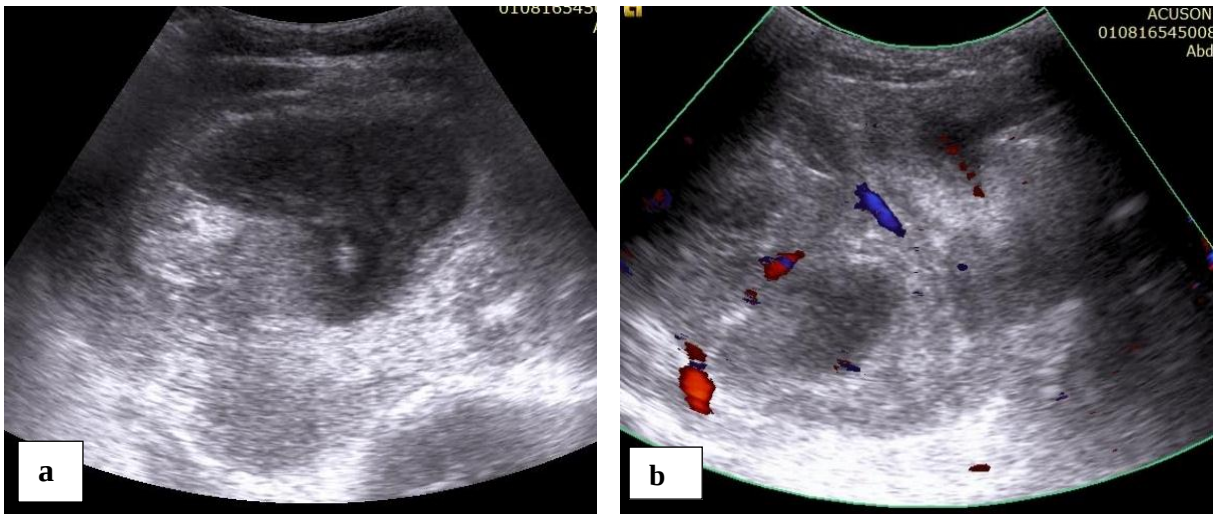

Figure 1: Ultrasound in axial section showing a heterogeneous pelvic mass(a); with color doppler signal (b).

An abdominal and pelvic CT-scan non-enhanced showed that the mass corresponded to the right pelvic kidney (Figure 2a) associated with significant hydronephrosis. Pyelic cavity was measured at 65mm contrasting to a thin ureter. Final diagnosis was a giant hydronephrosis in pelvic kidney with uretero-pelvic junction obstruction (Figure 2b).

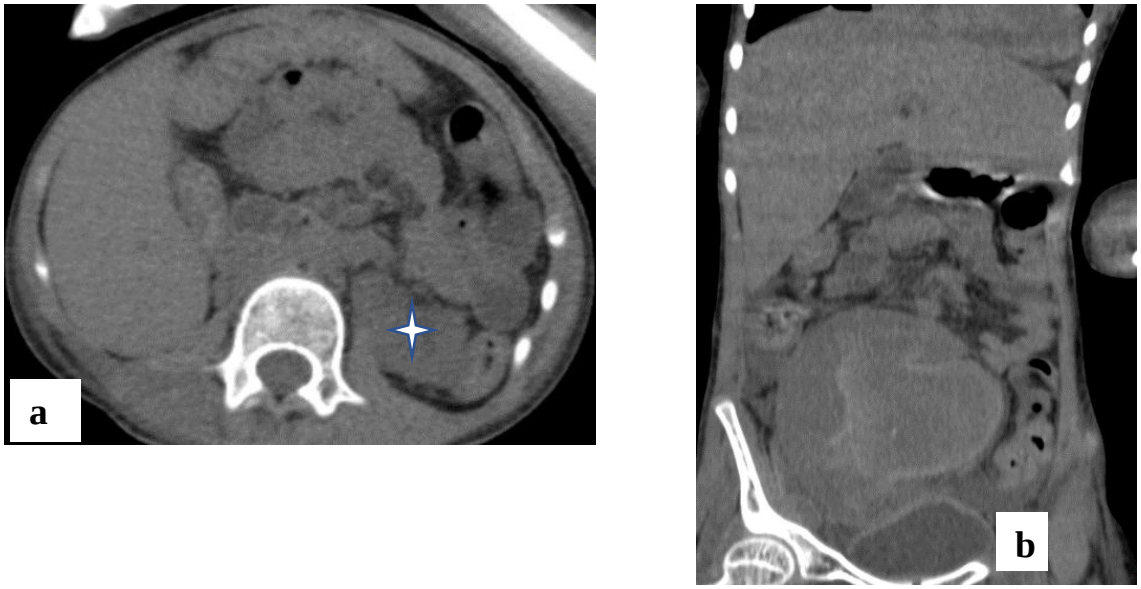

Figure 2: Abdominal CT–scan non enhanced in axial section showing the empty right renal compartment, star-shaped on the left: left kidney (a); coronal reconstruction through the pelvic region showing the pelvic ectopic kidney increased in size with giant hydronephrosis (b).

The child received medical treatment for the urinary tract infection and percutaneous nephrostomy. The evolution at 3 months was good with a normalization of the renal assessment and complete disappearance of the symptoms.

DISCUSSION

The pelvic ectopic kidney is defined by the location of the kidney below a horizontal plane passing through the iliac crests.[3] Its incidence varies from one country to another, rangin g from 1/5000 to 1/12000.[1,4] Embryologically, the kidney begins to develop in the pelvis and then migrates to the lumbar region around the 9th week of amenorrhea. This migration is facilitated by the caudal growth of the embryo. On the one hand, during the lumbar ascent of the kidney, it acquires its blood supply from the external and internal iliac vessels and the aorta after the 8th week of amenorrhea. Therefore, an abnormality in the origin of the renal artery may be responsible for a kidney migration defect. On the other hand, several factors may interfere with renal development such as genetic factors, maternal metanephritic disease, poor ureteral bud development and teratogenic causes.[5] The pelvic kidney predominates in males with 65.7% of cases.[6]

Giant hydronephrosis is clinically defined as a dilation of the excretory tract containing at least 1 liter of fluid or the equivalent of 1 day's urine in children where fluid constitutes 2-4% of body weight.[7] Obstruction is due to uretero-pelvic junction syndrome as in our case. High

insertion of the ureters into the pelvic kidney is responsible of an delayed emptying may be the origin of that the ureteropelvic junction.[8] It is often accompanied by total destruction of the kidney. In the case report of Rabii A and team, it reached 8 liters of fluid.[9] In our case, the drain brought back 1.5 liters. The association of a pelvic kidney with giant hydronephrosis is rare. Compared to the kidney in the normal position, the pelvic ectopic kidney is not more susceptible to disease except for stone formation.[3]

The pelvic kidney is usually asymptomatic and is discovered by chance during radiology examination.[3,4] It can manifest itself as various clinical signs such as colic, urinary incontinence, urinary tract infections, and high blood pressure.[5] In the case series study conducted by Engelhardt PF, urinary tract infection was the most frequent presentation after the asymptomatic forms, with 7/17 cases.[6] In a similar study by Benchenkroun, pain was predominant and was present in all patients. In the same study, there was one case in a 55-year-old man in which a mass in the left iliac fossa was the circumstance in which the pelvic kidney was found.[3]

The pelvic kidney may be associated with other congenital abnormalities such as vaginal atresia or unperforated hymen, hypoplastic uterus, and rudimentary fallopian tubes and ovary.[10] Overall renal function in children with an ectopic pelvic kidney is generally normal. On nuclear analysis, the pelvic kidney shows a 2/3 reduction in function with a p-value of 0.002.[6] In adults, most patients diagnosed with ectopic pelvic kidney disease have had impaired renal function or even a silent kidney.[3]

Radiological investigations allow the diagnosis of a pelvic kidney and check associated pathologies. They appreciate also the contralateral kidney.[7]

Abdominal-pelvic ultrasonography is the first-line exploration. In children, ultrasound is usually sufficient to make the diagnosis by showing the position of the kidney below the iliac vessels.[6] The kidney is usually atrophic.[5] It is often on the left side.[6] Frequently identified associated urologic pathologies were lithiasis and junction syndrome. Almost one third of children with a pelvic kidney have concomitant pathology.[6]

Intravenous urography shows the situation of the kidney and the associated pathologies.[3] However, this examination is invasive and rarely indicated in children.

The abdominal-pelvic CT scan is indicated in bilateral forms to look for possible renal symphysis and for the assessment of the extension of a pelvic kidney tumour.[3] The ureter of the ectopic pelvic kidney is usually short. In our case, hydronephrosis was not associated with a thinning of the renal cortex despite its importance. In our case, the pelvic kidney was right-sided, hypertrophic and associated with giant hydronephrosis.

CONCLUSION

The pelvic ectopic kidney associated with giant hydronephrosis can simulate a pelvic tumour in the clinic as well as on ultrasound. The abdominal-pelvic CT-scan is necessary for this rare form in children, especially in front of a pelvic mass and non-visualization of the kidney in its normal anatomical position.

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