Cas clinique / Case report

Proximal Ureteral Quadruplication in Combination with Ureteral Cyst: a Case Report.

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Abstract

Ureteral quadruplication is a very rare developmental malformation and it is sometimes associated with other congenital urinary tract abnormalities. This is the first case described in our country and in Africa, and apparently the 13th worldwide. It concerns a 9 years old Black child of the male gender, with a history of distended and progressively painful abdomen. Six years ago, the patient was brought to the hospital for a painful abdominal mass associated with fever and cough. Fever and cough was finally found to belong to malaria and upper respiratory tract infection. In the CT-scan, the abdominal lump was reported to be a polycystic kidney in combination with ureterohydronephrosis. After a laparotomy indicated for this former condition, per operative diagnosis was that of a unilateral quadruplication of the right ureter, opened into a close proximal ureteral cyst. The distal monoureter opened into the bladder. Surgery of this malformation was conservative. It consisted of ureteral resection of its dilated proximal portion, and nephro-ureteral end to end anastomosis after ureteropyeloplasty. Post-operative outcome was marked by the fact that the child had clinically recovered from the previous symptoms and his paraclinic exams and laboratory tests were progressively satisfactory. In conclusion, in the presence of an abdominal mass associated with urinary tract signs in late childhood, the physician should also evoke ureteral malformation in his differential diagnosis. In terms of differential diagnosis, this case has a broad clinical impact across medicine because it is implicating both surgery (urology and pediatric surgery) and internal medicine (nephrology and medical oncology). In the other hand, renal conservative surgery is to be considered when the secretory function remains.

Key words: Ureter, quadruplication, ureteral cyst, nephropyeloplasty, vesico-ureteral reflux.

Résumé

La quadruplication rénale reste un fait clinique rarissime. Les auteurs rapportent le cas d'un enfant âgé de 9 ans, amené en consultation pour une masse para ombilicale et du flanc droit, dans un contexte de toux fébrile. Initialement étiquetée à l'échographie des voies urinaires comme une polykystose rénale droite avec en différentiel l'hypothèse d'une hydronéphrose sur syndrome de la jonction pyélo-urétérale, cette masse va s'avérer correspondre à une ectasie de la portion initiale de l'uretère droit, associée à une quadruplication rénale et pyélique. Le traitement chirurgical a consisté en une néphro-pyéloplastie droite assurant la confluence des quatre pyélons dans un néo-bassinet unifié, suivie d'une anastomose pyélo-urétérale, après urétérectomie partielle emportant l'ectasie. Malgré une hydronéphrose droite résiduelle, les suites opératoires à trois mois de la chirurgie sont bonnes, caractérisées par une stabilité clinique et biologique.

Mots clés : uretère, quadruplication, kyste urétéral, néphropyéloplastie, reflux vésico-urétéral.

INTRODUCTION

Among renal and ureteral multiplicities, the quadruplication is the rarest malformation [1-4]. In the early year 1994 for example, there were only 3 cases known in the world and all of them concerned adult patients [4]. Since that time, it has been showed that children too present that malformation [4-8]. This malformation There other abnormalities associated, including when the malformation is bilateral [7-9], massive vesico-ureteral reflux [10], dystrophic kidney [11] and proximal blind-ending branches of the quadruplicated ureter [12]. The present case report concerns a Black race child of male gender, who came to the surgical consultation for distended and painful abdomen. The physical examination palpated abdominal mass, located from the umbilicus and right lumbar region to the pelvis, which was initially diagnosed as a right polycystic kidney by abdominal CT-scan. The aim of this report is to present the diagnostic difficulties, describe the surgical technique used and the very good post operative outcome and to discuss about therapeutic

CASE PRESENTATION

A nine years old patient of the male gender, born at term, with normal psychomotor development and doing well at school was brought to consultation six years ago (31/05/2007). This child of the Black race is originating from the center region of Cameroon in Central Africa and his parents are Roman Catholics. His vaccines were up to date and his known blood group is O rhesus positive. He was received at pediatric surgery consultation for a painful and compressive abdominal mass extending from the umbilical region and the right flank to the hypogastric region of two weeks duration. According to the history, the abdominal lump was intermittent, occurring from time to time since the infancy, with associated pollakiuria, painful micturition. There was an unset of a productive cough, fever and constipation at the time of consultation. On physical examination, the child had a good general state, weighing 24 kg. His temperature was 39oC and a pulse rate of 130 beats per minute. Crepitations were noted in base both lung fields. The abdomen was deformed, with a lump projecting at umbilical and hypogastric regions, extending to the right flank. The lump was renitent, mobile, not tender, dull and no murmurs (figure 1).

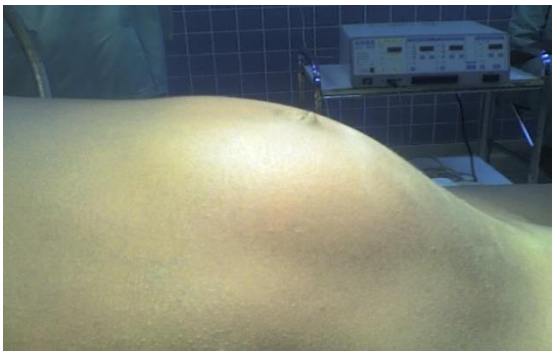

Fig 1: abdominal lump

There were no lumbar contact and rectal examination was normal. Laboratory workup with full blood count showed leucocytosis with polymorphic. The blood film also identified Plasmodium falciparum. The renal function test showed increased blood urea and creatinine but with normal creatinine clearance. Bacteriologic examination of urine was normal.
A chest X-ray showed shadow both basal lung fields. An ultrasound of urinary tree presented a polycystic right kidney with hydronephrosis due to pyeloureteral junction syndrome as differential. An abdominal CT scan revealed of multilocular cystic mass of the right kidney which was functional (figures 2).

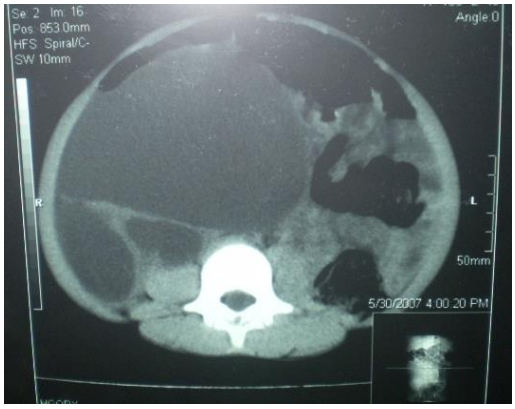

Fig 2: abdominal CT scan, right renal multiloculated cystic renal mass.

Intravenous pyelogram showed distention of the first portion of the right ureter, pyelorenal dilatation with grade 3 hydronephrosis. The clinical picture was then that of dilated upper urinary tree due to a nearby obstruction associated to febrile lung infection and malaria in an endemic zone. The medical indication was double aimed at treating the medical condition of lung infection and malaria and upper urinary tree obstruction by surgery. The preoperative workup, consisting of fasting blood sugar, plasma proteins,

blood electrolytes, coagulation and bleeding times were normal. The child is of the blood group O rhesus positive, the hemoglobin electrophoresis is type AA.

On admission, he was treated for malaria with intra-muscular artemesine 80 mg daily for 3 days, lung infection with cefuroxime 200 mg per os 12 hourly for 10 days, carbocystein suspension given orally for 5 days. This treatment enabled the patient to improve within 4 days; the immediate preoperative preparation consisting of fasting from the eve of surgery. The operation was conducted 13 days after his admission. The patient was supine under general anesthesia and endotracheal intubation, sandbags on the flanks. A nasogastric tube and transurethral catheter were in place. The pediatric surgical team were re-enforced with an urologist. An extended right umbilical transverse incision was made, revealing a right renal and pyelic quadruplication, corresponding to the palpable abdominal lump. The four calices were hydronephrotic, emptying in a single ureter, whose proximal portion was saccularly dilated (figure 3). It was however noted that this malformed kidney was quite functional. The left kidney and urinary tree were normal. Since the 4 calices were functional, the surgery consisted of a removal of the sacculated ureteral cyst and nephropyeloplasty assuring continuity of the calices into a single pelvis.

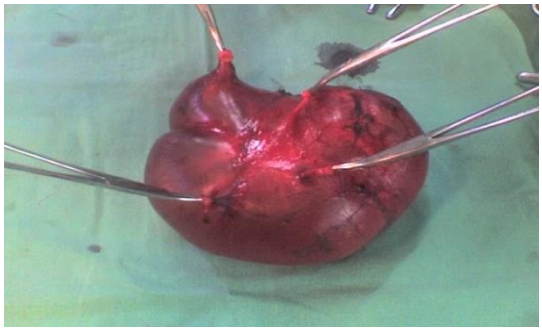

Fig 3: sacculated ureteral cyst with the four ureters.

This was followed by pyeloureteral anastomosis after resection the dilated proximal ureteral portion. The end to end pyeloureteral anastomosis was protected by an ureteral stent through the new pelvis, and brought out through a stab wound on the right flank just below the transcutaneous nephrostomy by a Foley catheter (figure 4). The retroperitoneal cavity was drained with a suction drain.

The operation lasted 3 hours 5 minutes and the child received 2 units of blood.

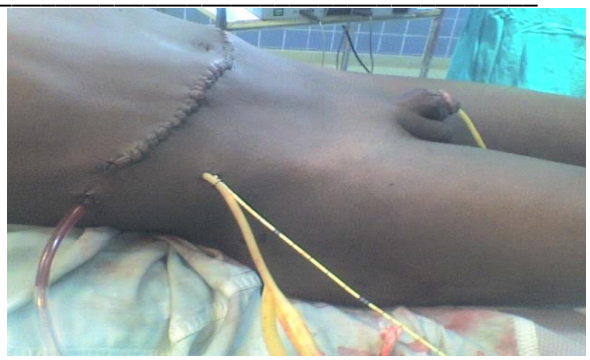

Fig 4: post operative picture; catheter and drains in place.

Post operative treatment consisted of infusions of crystalloids 1000 ml daily. The antibiotics consisting of cefuroxime 500 mg 8 hourly and metronidazole 500 mg 8 hourly were given for 5 more days. He also received painkillers consisting of subcutaneous tremadol 50 mg 8 hourly and paracetamol 500 mg 12 hourly slow I.V. Post operative monitoring consisted of hemodynamic state, bowel motion, symptoms and signs of renal function and urogenital infection. The post operative evolution was satisfactory and on the 5th post-operative day the nephrostomy was clamped and retroperitoneal drain removed. The other drains and catheters were removed on the 9th day. A control intravenous pyelogram done on the 12th post-operative day showed a residual vesico-ureteral reflux which ended by itself after a year. The patient was discharged on day 16. At post operative follow-up, the evolution was satisfactory, with a good general state, no urinary symptoms until nowadays 3 years later. Seventeenth months after the surgery, there is no symptom and the physical examination of the boy is quite normal. The biological markers of renal function have returned to normal values since the third month following the plastic surgery. A test of the renal function with a scintigraphy which was expected was satisfactory. Other lab tests are expected in each follow-up and the patient is now seen every twelve months.

DISCUSSION

Nothing but the operative exploration found the diagnosis of ureteral quadruplication. The pre operative presumptive diagnoses we evoked were polycystic right kidney probably due to pyeloureteric junction syndrome. From this patient history, painful mictiuration was the only urologic initial symptom, followed by progressive abdominal mass which accompanied an infectious syndrome. This explains why the diagnosis of the malformation could not be evoked clinically. Even with a CT scan and an intravenous pyelogram done, this developmental

abnormality of the upper right ureter was not obvious. Actually, because of its rarity, very few cases of ureteral quadruplication have been reported in the World till date [1-12]. Clinically, in addition to the sensation of abdominal mass or abdominal discomfort, patients with such a malformation also present with a history of repeated urinary tract infections [6, 8-10]. Ureteral quadruplication is always associated with ureteral cyst and vesico-ureteral reflux. The later condition is explained by irregular abundant urinary inputs, which lead to failure of the ureteral peristaltic motion as formerly suggested by Klinge and al [6]. As the pre-operative tests results and intraoperative findings showed a satisfactory secretive function of the kidney, we performed a pyeloplasty by opening and suturing the four remaining portions of the quadruplicated ureter, followed by termino-terminal pyelorenal anastomosis. In most cases, nephrouretrectomy is the surgical indication, since in these cases there is no more renal excretive function [7, 8, 10-12]. Although the renal function of our patient has returned to the normal in the early controls, there was a post-operative residual vesico-ureteral reflux which persisted for a year. In the opposite, Lopez and al defends to systematically remove the apparently normal kidney which has been suffering from vesico-ureteral reflux by laparoscopy. They argue for a poor prognostic functional efficiency of such a kidney [7]. In case of poor or absent renal function, nephrouretrectomy is the justified radical cure. When the afflicted kidney is still functioning, conservative surgery is to be considered as we did and is defendable. Although there is a residual vesico-ureteral reflux, the secretive renal function has recovered and is better 6 years after the surgical repair. The case is still under routine medical surveillance.

CONCLUSION

This case of ureteral quadruplication in a pre-teen age show how this abnormality of development is not life-threatening in infancy, but is associated with a burden in terms of urinary tract repetitive infections and abdominal discomfort linked to the urinary mass. According to the diagnostic aspect, this case has a broad clinical impact across medicine because it is implicating both surgery (urology and pediatric surgery) and internal medicine (nephrology and medical oncology). In the presence of an abdominal mass associated with urinary tract signs in late childhood, the physician should also evoke ureteral malformation in his differential diagnosis. With the kidney well conserved, the conservative option was our choice. This therapeutic option is to be considered while discussing surgical indications.

CONFLICTS OF INTEREST:

The authors declare that they have no competing interest.

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AUTHORS' CONTRIBUTIONS

JMB consulted, treated and performed the surgery in a team including an urologist and a general surgeon. As urologist, PJF orientated and performed as advisor in the nephropyeloplasty. DA was a major contributor in writing the manuscript. Histopathology examination of the surgical piece was performed by JMMN. From French, the translation of the initial text was realized by PMN. All the authors read and approved the final manuscript.

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