

Congenital Hydroureteronephrosis in a 5-Year-Old Male Patient: An Unusual Presentation

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ABSTRACT

Background: Congenital Mid Ureteral Stricture (CMS) on a 5 year old is relatively uncommon, particularly without previous symptomatology.

Case presentation: A 5-year-old Mexican boy presented with right abdominal pain 1 month before surgery. He was diagnosed of hydronephrosis by ultrasound, being ureteropelvic junction obstruction the suspected diagnosis. The patient was submitted to pyelography before a laparoscopic surgery done, finding multiple adhesions at the mid ureteral area causing a stricture, it was resolved immediately by ureteroureterostomy and a double pigtail stent. Patient was discharged from the hospital 3 day after surgery. He had no apparent sequelae and by the time hydronephrosis is lower than then, after 3 months follow up.

Conclusion: Congenital Mid ureteral stricture is not a common pathology that needs to be reached by the urologist; not diagnosing this may lead to renal atrophy and malfunction. We conclude that, although rare, unilateral hydronephrosis in a child should be recognized, immediately diagnosed and treated.

INTRODUCTION

Background: Hydronephrosis is a common phenomenon. In most of the cases (63 %) prenatal hydronephrosis can be associated with normal renal physiology. This could spontaneously return to normal in the first year of life [1], just requiring follow-up. In less cases is a severe condition requiring surgical approach and sometime with urgency. The most frequent causes of hydronephrosis requiring surgery are ureteropelvic junction obstruction (11%), vesicoureteral reflux (9 %) and vesicoureteral junction obstruction (4%) [2]. By definition the renal pelvis become distended with urine because the ureteropelvic junction cannot conduct it properly, Congenital Mid Ureteral Stricture (CMS) is a rare cause and its typically diagnosed intraoperatively [3]. Characterization of stricture site is important to plan the best surgical approach, and, whether it's urgency or not [4]. Here we report a case that was managed in our institute.

Case presentation: At the age of five, a male child presented with right abdominal pain in emergency room. He got an abdominal ultrasound done, a right hydronephrosis was detected; the left kidney appeared to be normal (Figure 1).

A Voiding Cysto-Urethrogram (VCUG) produced normal findings. Subsequently, a MAG3 renogram revealed a hydronephrotic obstructed right kidney with reduced

global cortical uptake, without response to diuretic even after 30 minutes, and renal function of 38% (Figure 2).

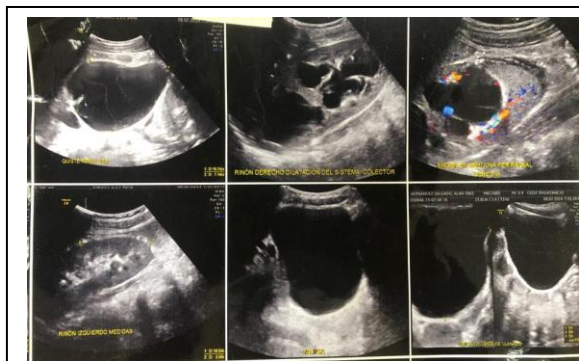


Figure 1: Renal ultrasound with right hydronephrosis.

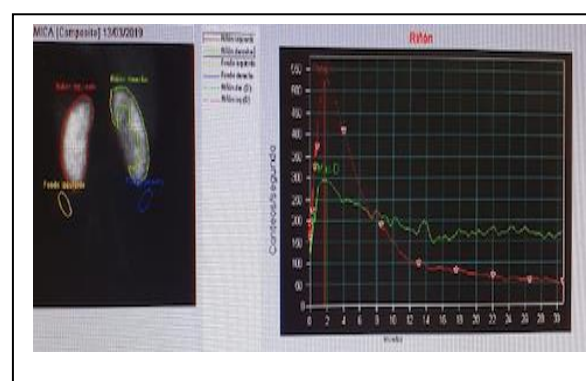


Figure 2: MAG-3 renogram with reduced global cortical uptake, without response to diuretic and apparent normal left kidney.

The patient was admitted electively and underwent a Retrograde Pyelography (RGP) in the operating room. Right CMS was detected by retrograde pyelography (Figure 3). Therefore we decided a surgical intervention by laparoscopy. An 8 Fr. Foley catheter was placed for bladder drainage, the patient is then placed in a modified right lateral decubitus position, with the thorax rotated back slightly at 30 degrees and both legs flexed by the knee 90 degrees, incision is made and the optical trocar is placed at the umbilicus by Hasson technique, then we place under visual guidance other two 5mm trocars through the para-rectus line so that all ports are distanced by at least 2-3 cm, then we dissect the peritoneum and spot the stricture (Figure 4). It was resected (1cm), and ureteroureterostomy was done, no adhesion or external mass was found, we decided to place a double-J stent and close with non interrupted 4-0 monocryl suture. A pathological review revealed focal chronic ureteral inflammation with narrow lumen

(1 mm). Stent was removed at 6 weeks of the procedure, showing a better lumen in RGP (Figure 5). It has been six months of follow up of this patient; no pain or infection has been reported at the time.



Figure 5: Right retrograde pyelography after 2 months of surgical treatment.

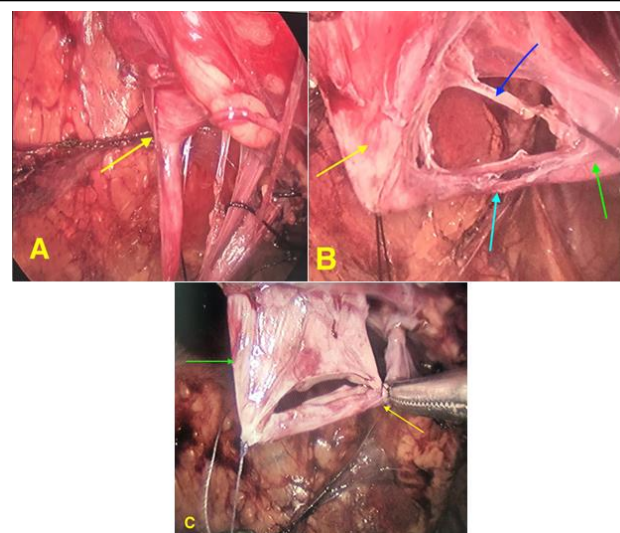


Figure 4: CMS seen in laparoscopy. A. CMS shown on yellow arrow, the proximal ureter being dilated. B. yellow arrow showing proximal ureter, light blue arrow showing ureteral stricture, green arrow showing distal ureter, blue arrow showing ureteral vessels. C. Ureteral anastomosis, yellow arrow showing distal ureter, green arrow showing proximal ureter.



Figure 5: Right retrograde pyelography after 2 months of surgical treatment.

DISCUSSION AND CONCLUSION

CMS is a rare cause of hydronephrosis and is much less frequent than proximal or distal stricture [5]. Back in 1937 Campbell published an autopsy series of 12 thousand children. There he found congenital ureteral obstruction in 1:150 autopsies. Only 4% of them had a CMS [6]. There are many theories to explain stricture formation during embryogenesis, including a localized area of developmental arrest caused by extrinsic compression by fetal vessels during intrauterine life, a congenital ureteral valve, intrauterine ureteritis, multiple infections and incomplete recanalization of the ureter [3,7,8]. The exact explanation remains unclear. CMS may appear as a definite stricture or as a true valve without lumen stenosis [9]. In the case presented, there was a complete lumen stenosis. CMS is typically not diagnosed preoperatively, and definite diagnoses have been reached via retrograde imaging of the ureter [5,10]. RGP remains controversial as a routine preoperative imaging procedure in cases of congenital hydronephrosis. Routine preoperative RGP is recommended in cases involving a diagnosis of an unexpected ureteral lesion, such as mid-ureteral stricture, ureteral polyp and retrocavalureter [8]. Hawang et al., recommended routine retrograde pyelography prior to repair, during the same surgical event, unless the ureter distal to the point of obstruction

has been well visualized by other means [4]. On the other hand, Rushton et al., did not recommend routine RGP based on findings from 108 pyeloplasties performed between 1986 and 1992, they found that RGP was not necessary for successful repair [10]. Although Grattan et al., [11], suggests the use of preoperative magnetic resonance, it is still unclear in diagnosing a CMS, and we strongly recommend performing preoperative RGP during the same anesthetic event, when neither the ultrasound nor MAG-3 renogram is clear about the cause of hydronephrosis (Figure 3). The management of CMS involves resection of the stricture and reanastomosis of the ureter, should never be treated conservative [5]. In our management, we decided to follow all the literature, and resected the stenotic area and re-anastomose the ureter in an spatulated fashion; it was all made by laparoscopy (Figure 4). Postoperative follow-up (6 months) revealed the decrease in the grade of hydronephrosis from severe to mid grade. In most of the relevant literature, improvement in hydronephrosis and renal function has been reported during short-term follow-up. In conclusion CMS is a rare cause of congenital hydronephrosis that should be considered whenever proximal mega-ureter is an associated finding. Urologist should be aware of it and patient should get proper surgical treatment. There are a lot of radiological modalities to make diagnosis and take decisions but retrograde pyelography remains one of the best options to diagnose ureteral anomalies.

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