



Congenital stenosis of the proximal ureter (A case report)

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ABSTRACT:

Congenital ureteral stenosis is discovered at autopsy in about 0.6% of children. Stenosis are usually an exaggeration of normal ureteral narrowing and are usually encountered in regions of physiologic narrowing of the ureters.

We report a case of a 6-month-old infant who presented with pyeloureteral junction syndrome with a dilated renal pelvis measuring 50mm, and intraoperative, we discovered a stenosis of the proximal ureter. The stenosis extended over 3cm and was located 15mm from the pyelocaliceal junction. We performed a pyeloplasty using the Anderson Hynes technique.

Congenital ureteral narrowing is a microscopic diagnosis, characterized by a relative reduction of the muscular mass of the affected ureteral segment, without an increase in fibrous tissue and with normal transitional epithelium.

There are four main causes of ureteral narrowing: congenital, inflammatory, surgical, and tumor-related.

Surgical management involves resection of the stenosis and uretero-ureteral anastomosis or bypassing the stenosis using a prosthesis, ileum, or appendix.

Key words: Ureteral stenosis, congenital, Anderson hynes, Pyeloplasty

I. INTRODUCTION:

Urinary strictures, whether congenital, inflammatory, iatrogenic, or tumoral, present a difficult problem for pediatric urologists. These obstructions, which are often found at locations where the ureter naturally narrows, such the pyeloureteral junction, call for specialized surgical care that is particular for each patient.

In this work, we are going to study a specific case of acute pyelonephritis that resulted in congenital ureteral stenosis in a 6-month-old infant. We will explore the surgical techniques used, the therapeutic strategy, and the post-operative issues.

II. OBSERVATION:

With the exception of an episode of acute pyelonephritis that necessitated radiological evaluation, the 6-month-old infant had no specific medical history. A radiological examination showed pyelocalyceal dilatation, suggesting a pyelocaliceal junction syndrome, with anteroposterior renal pelvis diameter of 50 mm and lamellar cortex.

Clinical examination revealed the infant to be in good general health, with appropriate development in terms of stature, weight, and mobility.

Laboratory test results demonstrated stable renal function with a creatinine level of 4.29 mg/l and urea level of 0.24 g/l.

In the operating room, the patient was placed under general anesthesia, intubated, and ventilated in left lateral decubitus position. An incision was made midway between the 12th rib and the iliac crest. The retroperitoneal region revealed a dilated pelvis and proximal ureter dilatation upstream of a stenosis, located 15mm from the pyeloureteral junction, extending over 3 cm and transitioning to a narrow ureter, suggestive of a congenital stenosis of the proximal ureter. Resection of the stenosis portion was performed using a pyeloureteral anastomosis according to the Anderson Hynes technique, with placement of a double J stent.

The procedure was straightforward, and the patient was discharged on postoperative day 10.

At 30 days post-op, ultrasound showed reduction of hydronephrosis with an anteroposterior renal pelvis diameter of 30 mm.

Removal of the double J stent was performed after 2 months.

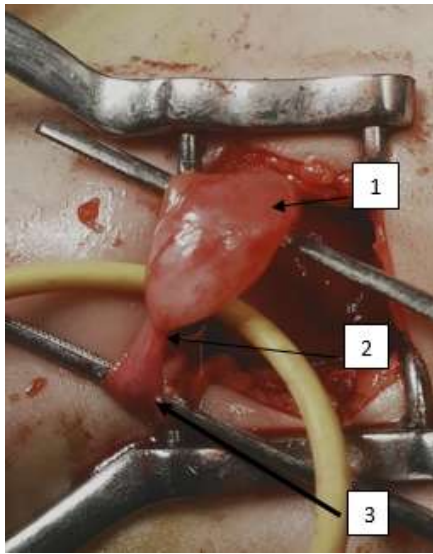


Figure 1

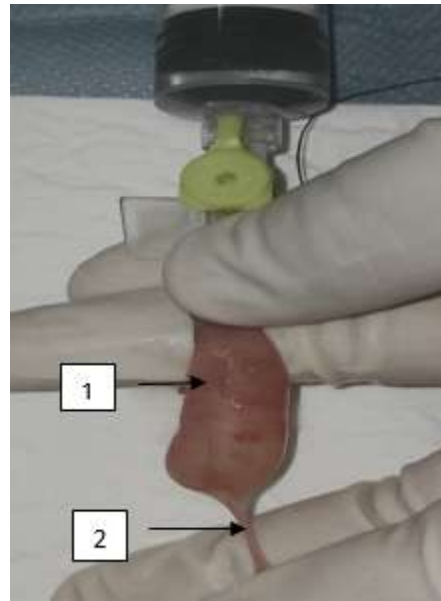


Figure 4

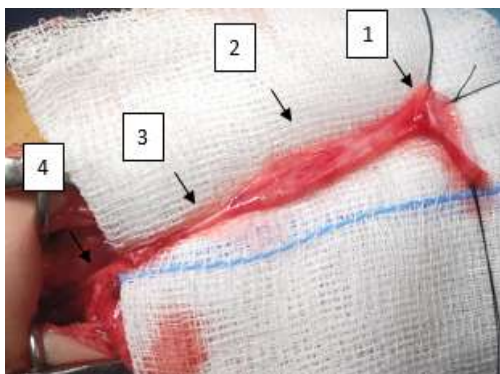


Figure 2

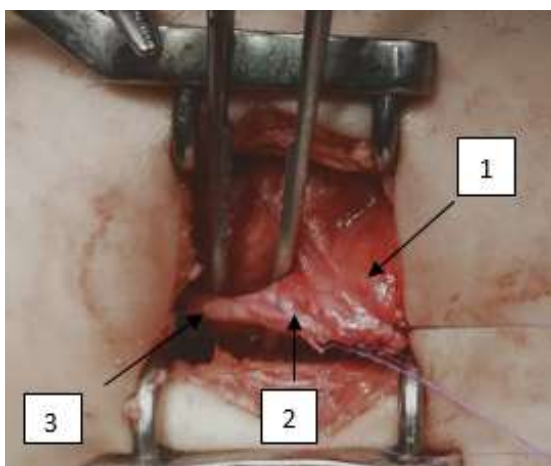


Figure 3

III. DISCUSSION:

Various factors can lead to ureteral stenosis: inflammatory, iatrogenic, congenital, or tumoral. [1] Most commonly, congenital stenosis occurs in areas of ureteral narrowing: the vesicoureteral junction, the pyeloureteral junction. [2]

According to Allen, these strictures are caused by a delay in embryogenesis at the 12th week of gestation due to vascular compression occurring at that time. [3]

A number of factors, including location (proximal, middle, or distal ureter), extent, and number influence urinary stenosis correction. For proximal strictures, ureteral resection – reimplantation is advised; for middle ureteral strictures, terminal uretero-ureteral anastomosis-resection; and for distal strictures, pyeloureteral anastomosis-resection is advised. It is also advised to use endoscopic techniques like ureterotomy or permanent endoprosthesis. [4]

The real difficulty for the surgeon is in treating large, complex strictures. In fact, ureteric repair requires surgery employing appendiceal or ileal interposition, as well as grafts from the bladder and oral mucosa. [4]

Melnikoff first described the use of the appendix in managing ureteral stenosis in 1912. [5] This technique is primarily recommended for strictures extending over more than 2 cm. The use of the appendix is more effective than that of the ileum due to the caliber of the appendix, which allows for anastomosis without requiring remodeling, it is pelvic position, and the absence of absorption function. [1] As for ileal interposition, it



presents complications such as hydro-electrolytic problems, strictures caused by intestinal mucosal secretions, absorption issues, and urinary tract infections. [4]

Some authors suggest using oral mucosa to reconstruct the stenosis ureter, especially for the proximal and middle forms. [4]

IV. CONCLUSION:

In conclusion, to treat congenital ureteral strictures in infants, a precise and personalized surgical approach is necessary to restore ureteral permeability while preserving renal function. Recent advances in surgery offer promising options, but close collaboration between specialists is crucial to achieving optimal outcomes. Further studies are imperative to improve diagnostic, treatment, and long-term follow-up methods for this complex clinical condition.

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Figure legend:

Figure 1: Intraoperative image: 1: Dilated proximal ureter; 2: Zone of disparity in caliber; 3: Normal ureter.

Figure 2: Intraoperative image 1: Pyelon; 2: Dilated proximal ureter; 3: Area of stenosis; 4: Normal ureter

Figure 3:Pyeloureteral anastomosis after resection of the stenotic zone 1: Pyelon; 2: Pyeloureteral anastomosis; 3: Ureter

Figure 4: Surgical specimen with dilated proximal ureter and zone of stenosis showing tight stenosis not allowing saline passage 1: dilated proximal ureter; 2: zone of stenosis