Bilateral congenital midureteric stricture

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Citation

Abstract
Midureteric stricture is a rare cause of congenital hydronephrosis. Its etiology and pathophysiology is uncertain. We report a case of an 8 year old male child who presented with recurrent urinary tract infections and moderate renal impairment. Sonar revealed bilateral hydronephrosis. This was confirmed to be due to intrinsic midureteric narrowing on MRU. Bilateral ureteric exploration confirmed these findings and uretero-ureterostomy was performed. Postoperative imaging showed improvement in hydronephrosis. We review the published literature.

CASE REPORT
An 8 year old boy was admitted to our institution with a documented UTI. He had no significant past medical history. Physical examination was non-contributory. He was in moderate renal impairment with a serum Creatinine of 130mmol/l and an estimated Creatinine Clearance, using the Schwartz formula, of 50ml/min/m?. Urine cultures for Tuberculosis were negative.

Renal sonar revealed gross bilateral hydroureteronephrosis with thin renal cortices. The bladder appeared normal. Of note no ureteric dilatation was seen at the level of the bladder.

An MCUG revealed a normal bladder and posterior urethra with no reflux. Radioisotopic mercaptoacetyltriglycine (MAG3) renography showed poor uptake bilaterally with apparent holdup at the midureteric level.

In view of the unusual imaging and to exclude extrinsic retroperitoneal pathology a Gadolinium enhanced magnetic resonance urogram was requested. This showed no retroperitoneal pathology, but bilateral midureteric obstruction. (See Figure 1).

Figure 1
Figure 1: Gadolinium enhanced magnetic resonance urogram demonstrating midureteric obstruction.

Retrograde pyelograms revealed a tight, short-segment, non-negotiable stricture at the level of the pelvic brim bilaterally. (Figure 2)
**Figure 2**

Figure 2: Left retrograde pyelogram showed a pelvic brim stricture.

Bilateral extraperitoneal ureteric exploration was performed using a Gibson incision. On both sides a similar appearance was seen. The stricture was excised revealing a pin-hole lumen. No valves or polyps were noted intraluminally. Stented ureter-ureterostomy was performed without tapering.

The child made an uneventful recovery. Followup sonar at 3 months showed improvement in hydronephrosis bilaterally.

Pathological review revealed proximal dilatation with nonspecific thickening of the muscular wall. Electron microscope evaluation of the strictured area showed no specific ultrastructural abnormality.

**DISCUSSION**

Congenital midureteric stricture is an uncommon cause of hydronephrosis in children. Less than 20 pathologically confirmed cases are reported in the literature. (1)

The diagnosis may be confused with ureteropelvic or ureterovesical junction obstruction. Hence, retrograde pyelography is essential in the management of this anomaly. The differential diagnosis must additionally include ureteral valves and fibroepithelial polyps.

Ectopic ureter and contralateral renal agenesis have been associated with some of the published cases. (2,4) The bilateral nature of our case would lend support to Kropp's assertion that contralateral agenesis and bilaterality point to the manifestation of a similar pathological process in this condition. (1)

The cause of congenital midureteric stricture, however, remains unknown. Docimo postulated a variety of etiologic possibilities: 1) improper ureteric recanalisation, 2) insufficient vascular supply, 3) persistent ureteric folds, 4) ureteric bud anomalies. (3)

It has been postulated that this condition runs a more aggressive natural history than other causes of congenital hydronephrosis. (1) This feature is born out by our case where longstanding obstruction resulted in renal impairment.

**References**

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