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Cross-Fused Ectopia with Proximal Ureteral Atresia

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Description

A very uncommon congenital defect connected to dysplastic kidneys is ureteral atresia. It is uncommon for ureteral atresia to coexist with other urinary abnormalities. The atresia may be short or long, unilateral or bilateral. Any ureter segment can be damaged, but the distal one is usually the one that is. Even less common is proximal ureteral atresia.

The ureter develops from a ureteric bud that extends from the met nephric mesoderm into the caudal end of the mesonephric duct (Wolffian duct) (renal blastema). Initiation of the ureteric bud occurs 28 days after formation. The development of the whole renal collecting system is induced by cephalad expansion and interaction between the ureteric bud and metanephric mesoderm. A portion of the mesonephric duct connecting the ureteric bud origin to the future bladder is absorbed into the bladder, creating the ureteric link with the bladder.

The ureteric bud is the origin of each kidney's ureter, renal pelvis, calyces, and collecting duct. Ureteral atresia and kidney dysplasia are the outcomes of the ureteric bud failing to form. According to a theory, ureteral atresia is brought on by an ischemia injury that occurs when the ureteral bud enlarges or when the growing kidney migrates, with the regional blood supply to the ureter shifting as a result [1].

The failure to canalise a portion of the ureter during the elongation of the ureteric bud may be the cause of the atresia. After the 40th day, canalization begins at the midportion and progresses to the cranium and cauda. Only 3 cases of proximal atresia and 14 cases of ureteral atresia have been recorded thus far, according to a survey of the literature. Rare and frequently accompanied by dysplastic kidney, ureteral atresia. Distal atresia with ipsilateral pelviureteric junction blockage has been recorded, despite the fact that ureteral atresia is rarely linked to other congenital defects [2].

In our case, the crossing fused ectopia with hydronephrosis that was not previously documented was connected to the ureteral atresia. The failure of the ureteric bud to form results in the development of multicystic dysplastic kidney. The presence of such severe hydronephrosis in a crossed fused ectopia with ureteral atresia can be explained by abnormal blood flow and the subsequent partial or complete ischemia of the ureter. With normal renal parameters, unilateral ureteral atresia may go undiagnosed for the entirety of a person's life if it is asymptomatic.

When atresia is suspected, retrograde or antegrade pyelography may be used to make the diagnosis. Early surgical intervention helped preserve renal function in children with ureteral atresia who presented with urinoma at birth, according to cases reported by Hedden et al. and Zundel et al. [3, 4].

It was hypothesised that the growing kidney managed to avoid dysplasia thanks to the decompression that followed the rupture. Although non-functioning kidneys and ureteral atresia are related, function has been seen to return after ureteral continuity and unimpeded drainage have been established. Several methods, including pyeloplasty, ureteric reimplantation, ileal conduit, and psoas hitch, can be used to do this, depending on the length and diameter of the native ureters that are available and the distance between the pelvis and bladder.

Considering ureteral replacement depends on the anatomy. In 1912, Melnikoff used the appendix to replace the ureter for the first time. Since then, various conditions such as ureteral atresia, transitional cell carcinoma of the ureter, long ureteric strictures, retroperitoneal tumors, and traumatic ureter injury have been treated with appendix substitutes. [5, 6].

For a tension free anastomosis to bridge the ureteric deficiency, the length of the appendix and its vascular pedicle is crucial. The appendix was able to bridge the distance from the renal pelvis to the bladder in this instance because the left kidney was ectopic, cross-fused, and its pelvis was close to the bladder.

Conclusion

In order to preserve renal function, ureteral atresia, a rare congenital abnormality, needs to be highly suspect and treated right away. The similar size to the ureter, the existence of peristalsis, and a reduced electrolyte imbalance are benefits of employing the appendix as a ureteral substitute. The caecal cuff can be used in conjunction with the appendix when the ureter and appendix have different calibres. Stenosis, fistula risk, and anastomotic leak are the drawbacks.

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