Introduction

Congenital Ureterovesical Junction Obstruction (UVJO) may be observed during fetal age or any stage at the time of childhood. It is aimed in this report to present a male child with obstructive megaureter due to congenital UVJO. He was surgically treated with excision of narrowed distal ureter in addition to tapering ureteroplasty with ureteroneocystostomy. The topic is discussed with special reference to etiology of this rather rare entity under the light of relevant literature.

Case Presentation

A 27-months-old boy was admitted to our department with an antenatal history of right Hydroureteronephrosis (HUN). Laboratory tests were otherwise normal except signs of Urinary Tract Infection (UTI) including leucocyturia. The microbiological urinalysis was positive for UTI revealing E. coli 10⁵ CF µ/mL. Urinary ultrasonography revealed an enlargement of the right kidney, pelvis and a megaureter with tortuosity down to the urinary bladder. Scintigraphy using Di-Mercapto Succinic Acid (DMSA) and Di-Mercapto Pentetic Acid (DTPA) showed a dilated right kidney with reduced scintigraphic uptake revealing a differential renal function of 41% for the right and 59% for the left kidney with signs and findings showing UVJO. Magnetic Resonance Imaging (MRI) showed enlargement of the right kidney with marked reduction of the cortical thickness, an enlarged pelvis and huge dilatation of right ureter with tortuosity down to the bladder (Figure 1). Due to progressive worsening of the upper urinary tract function and repeated UTIs, surgical intervention became a necessity rather than of choice. Surgical intervention revealed a severely narrowing of the right distal ureter for 2 cm in accordance with UVJO resembling bird’s beak and the diameter of the ureter proximal to UVJO was measured nearly 4 cm (Figure 2). Excision of the distal...
The etiology of primary UVJO is unclear. The transportation of urine at the UVJ occurs by means of peristalsis of the circular outer and longitudinal inner layers of muscles [6]. It has been postulated that increase in collagen and hypertrophy of the circular muscle and atrophy of the inner muscle may be responsible for the deterioration of urine flow at the UVJ [7-11]. It has been hypothesized that, with a resemblance to the gastrointestinal system, urinary tract obstructive disorders might be related with the aplasia or hypoplasia of the Interstitial Cells of Cajal (ICC) [12]. In a recent study by Kart and co-workers, it was shown that the number of ICC was decreased in the UVJ of the patients with primary obstructive megaureters compared to the normal control group [6]. Histological analysis in our patient revealed fibrosis of tissues removed at surgery.

**Conclusion**

UVJO is a rare but important cause of HUN in children and may occur at any stage during childhood. It is commonly associated with UTIs and deterioration of upper urinary tract. Timely and accurate management of this entity is important for the preservation of upper urinary tract function. The health providers dealing with such kinds of patients should keep this anomaly in mind and a prompt pediatric surgical consultation is recommended and the patient should be treated accordingly.

**References**

