

Zinner's syndrome: unusual urological presentation in adolescent

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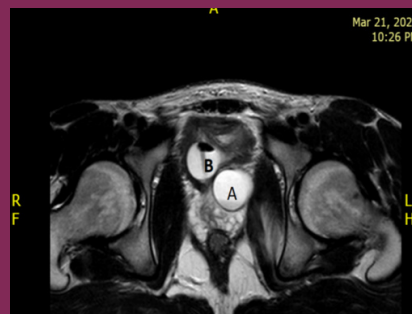
Introduction:

On 1914, Zinner's syndrome (ZN) was described as the classic triad of unilateral renal agenesis, ipsilateral ejaculatory duct obstruction, and ipsilateral seminal vesicle cyst. An incidence of approximately 0.00214 % in newborns was reported. Usually presentation was on fertility age group with ejaculatory or fertility concerns.

In our case, patient was adolescent. Presentation was atypical of this syndrome; he presented with acute urine retention.

Embryological:

The mesonephric or wolffian ducts are present in human embryos. In males, this duct is transformed under the control of the testosterone and the anti-müllerian hormone into the hemitrigone, the bladder neck, proximal urethra (up to the external sphincter, seminal vesicle, vas deferens, efferent ducts, epididymis, paradiidymis, appendix epididymis). Genetic disturbances (4-8 week) leads to distal mesonephric duct is improperly developed which leads to atresia of the ejaculatory duct which becomes an obstruction that leads to cystic enlargement of the seminal vesicle.

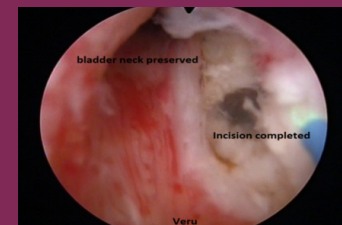
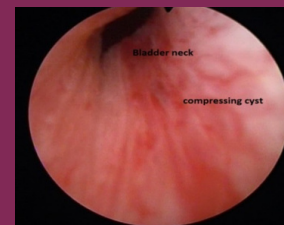


Case presentation:

Boy 17-years old presented with acute urine retention with left testicular pain. The condition was not associated with any constitutional symptoms. He had normal vital signs. On first assessment differential diagnosis by the family physician was aimed to exclude Torsion, trauma and severe UTI. On clinical examination the testis were normal and bladder was palpable on supra-pubic region. Later, laboratory tests did not support infection or any abnormalities including kidney function, inflammatory markers. Abdominal ultrasound showed the distended bladder and agenesis of left renal unit with a cystic swelling related to the base of the prostate compressing the bladder neck [picture-1]. Urinary catheter was inserted, with immediate passage of 600 cc of clear urine. Case was referred to urology for further assessment. Confirmatory MRI was done.

Surgical findings:

Diagnostic urethra-cystoscopy show normal anterior urethra. On prostatic urethra the lumen was deviated to right side and compressed with normal mucosa covering. It is compromised its diameter significantly. At the bladder neck there was a bulge on left side with hemi-trigon and right ureteric orifice was normally recognized. Using LASER fiber on cutting mode (Lumenis Pulse™ 120H Holmium Laser System), a small incision was created distal to bladder neck by approximate 1 cm and deepen gradually until cloudy content was expressed out.



Conclusion:

In the best of our knowledge- is first case report of urine retention as presenting symptom of ZN. Age presentation of the syndrome is always on fertile age group, and our case is the first diagnosed case on early adolescent. In addition to the Endoscopic urethral management and preserved ejaculatory function could relieve the obstruction of ejaculatory duct by time.