Zinner Syndrome Case Report - A Rare Developmental Anomaly of the Mesonephric Duct

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Abstract

Zinner syndrome is a rare congenital malformation characterized by the association of an ipsilateral seminal vesicle cyst, ipsilateral renal agenesis and ipsilateral ejaculatory duct obstruction due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct. Here we report a 19 year-old male patient presented with complaints of right groin pain radiating to the right testis and the medial aspect of the right thigh for the last 1 month. No history of lower urinary tract symptoms, painful ejaculation, hematuria, or trauma. General physical examination no abnormality detected. Ultrasound abdomen and pelvis shows absent right kidney with cystic structure behind the bladder. Computed tomography showed absence of right kidney noted in right renal fossa and along the line of its ascent. There is associated absence of ipsilateral renal artery and low lying right adrenal gland. A well defined cystic tubular lesion with bulbous anterior intra vesicle projection is seen cephalic and lateral to prostate along its right side seminal vesicle cyst which corresponds to Zinner syndrome. The patient was asymptomatic and was managed conservatively with analgesics during his follow-up in our clinic with disappearance of the pain. The Conservative management is the mainstay of treatment plan in asymptomatic patients. The patient was symptomatically better during his follow-up in our hospital with disappearance of the pain.

Introduction

Rare congenital malformation of urogenital tract associated with ipsilateral seminal vesicle cyst, ipsilateral renal agenesis and ipsilateral ejaculatory duct obstruction. Mainly due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct. It was first reported by Zinner in 1914 [1]. Till now only few hundreds of cases reported in the literature. Hereby we like to present about the rare developmental anomaly involving the Mullerian ducts encountered in our hospital [2,3].

Case Presentation

A 19-year-old male patient with no known comorbidities was came to our urology out-patient department with complaints of right dull aching groin pain radiating to the right testis and the medial aspect of the right thigh for the last 1 month. No history of lower urinary tract symptoms, painful ejaculation, hematuria, or trauma. Physical examination was unremarkable. Routine laboratory investigations and renal function tests done. Reports are within normal limits. Semen analysis was done within normal limits. Ultra Sound (US) abdomen and pelvis report from our hospital shows absent right kidney with cystic structure behind the bladder (Figure 1 and 2). Computed tomography showed absence of right kidney noted in right renal fossa and along the line of its ascent. There is associated absence of ipsilateral renal artery and low lying right adrenal gland. A well defined cystic tubular lesion with bulbous anterior intra vesicle projection is seen cephalic and lateral to prostate along its right side seminal vesicle cyst which corresponds to Zinner syndrome. The patient was asymptomatic and was managed conservatively with analgesics. During his follow-up in our hospital he is symptomatically improved with disappearance of the pain. The Conservative management is the mainstay of treatment plan in asymptomatic patients. The patient was symptomatically better during his follow-up in our hospital with disappearance of the pain.

Discussion

Rarest congenital abnormalities of the urogenital system which is usually present and diagnosed in the 2nd to 4th decade of life [3]. Incidence is 1 in 3000 to 1 in 4000 newborns. Patients with Zinner syndrome are usually normal but sometimes can present with decreased urine output, increased frequency, pain over the perineum, or epididymitis [3]. The Normal embryological development
of the genitourinary system begins at around 13th to 22nd week of gestation. Any disturbance during normal embryogenesis leads to this condition. It is a rare congenital disorder which includes other abnormalities like polycystic renal disorders, ipsilateral testicular agenesis, artesians of the vas deferens and Hemivertebra Radiological Imaging provides accurate diagnosis of the anomalies of the genitourinary tract and evaluation of Zinner syndrome includes X-ray KUB, Ultra Sonogram, Computed Tomography, and Magnetic Resonance Imaging. Ultra sonogram is a simple imaging technique which is used mainly in this condition to detect the absence of the ipsilateral kidney or to show cystic structures behind the bladder and to show anechoic structures [3-6]. CT findings might include an ipsilateral renal agenesis in addition to a well defined cystic tubular lesion with bulbous anterior intra vesicle projection is seen cephalic and lateral to prostate [3,7]. MRI is the imaging technique of choice in diagnosing this condition due to its high resolution properties in evaluating the seminal vesicles cysts and the ejaculatory ducts [3]. Seminal vesicles cysts treatment modality depends on the clinical presentation and appearance of symptoms, and in asymptomatic patients are usually managed conservatively with regular follow-up until they start to complain of symptoms. Surgical approach is needed only in symptomatic patients. Minimal invasive techniques like transurethral aspiration of cyst, excision of cyst or transurethral aspiration combined with Alcohol and Minocycline injection in the cyst, or transurethral cyst deroofing [8-11]. Infertility should be ruled out in male patients. The conservative management is the treatment of choice in asymptomatic patients.

**Conclusion**

The combination of ipsilateral renal agenesis and seminal vesicle cyst is quite unusual and one of the rarest urogenital tract anomalies. Simple imaging techniques like ultrasonography abdomen and pelvis will detect the rare urogenital tract anomalies. The Conservative management is the mainstay of treatment plan in asymptomatic patients with regular follow up. Surgical intervention needed in symptomatic individuals like surgical aspiration of cysts through perineal or laparoscopic approach or percutaneous cyst drainage. Our patient was diagnosed with Zinner syndrome incidentally by ultrasonography. The patient was asymptomatic and was managed conservatively with analgesics and advised regular follow-up.

**References**