ZINNER SYNDROME: RARE CASE REPORT

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Abstract

Zinner syndrome is the rare syndrome associated with congenital seminal vesicle cysts and ipsilateral upper urinary tract anomalies, such as multicystic dysplastic kidney. Congenital malformations of the seminal vesicle are uncommon and most of them are cystic malformations. If an insult occurs between 4th and 13th gestational week, the embryogenesis of the kidneys, ureters, seminal vesicles and vas deferens could be altered. Cysts of the seminal vesicle may appear with a mass effect, dysuria, epididymitis, or obstruction of the gastrointestinal and genitourinary tracts. Approx. 2/3rd of them are associated with ipsilateral renal agenesis, because both the ureteral buds and seminal vesicles originate from the mesonephric duct. They were first described by Zinner in 1914. Most patients are asymptomatic until the 3rd-4th decade of life. Till now only few hundreds of cases reported in the literature. Hereby we present the rare developmental anomaly involving the mullerian ducts encountered in our hospital.

Keywords: Seminal vesicle cysts, Ipsilateral renal agenesis, Mesonephric (wolffian duct)

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Case Report

A 31-year-old male patient with no known comorbidities, came to our radiology out-patient department with complaints of burning micturition & occasional hematuria since last 1 year. Physical examination was unremarkable. Routine laboratory investigations and renal function tests done. Reports were within normal limits. Semen analysis was done which is also within normal limits.

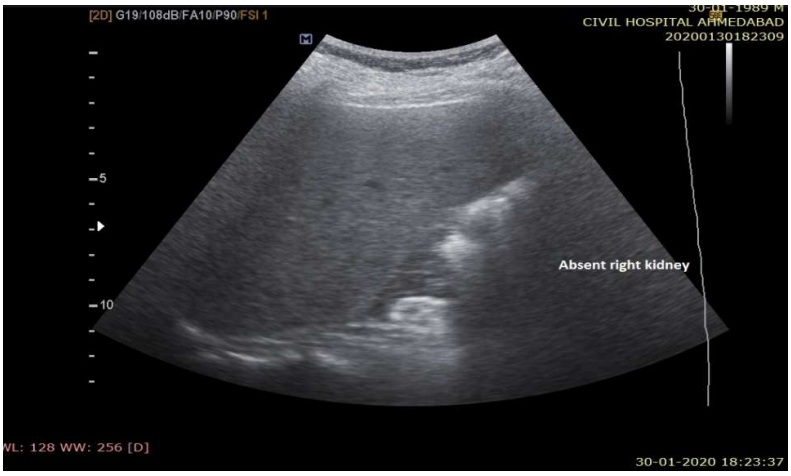
Ultrasound (USG) abdomen and pelvis report from our hospital shows absent right kidney with cystic structure behind the dome of urinary bladder. (figure 1). Computed tomography report shows absence of right kidney in right renal fossa and along the line of its ascent. There is associated

absence of ipsilateral renal artery and low lying flat right adrenal gland, revealed in arterial phase of CT scan study. (figure 2) A well-defined cystic tubular lesion with bulbous anterior intra vesicle projection is seen cephalic and lateral to prostate along its right side suggests possibility of seminal vesicle cyst. Rudimentary right lower ureter with ectopic insertion in above mentioned cyst was noted. (figure 3) On MRI correlation, seminal vesical cyst & rudimentary ureter appeared hyperintense on T1WI & hypointense on T2WI suggested possibility of proteinaceous retained secretion.

The patient was asymptomatic and was managed conservatively with analgesics. During his follow-up in our hospital he symptomatically improved with disappearance of the pain. The conservative management is the mainstay of treatment plan in asymptomatic patients.

Diagnosis of Zinner's Syndrome was made based on clinical presentation and radiographic findings.

Figure 1 USG of Abdomen & Pelvis



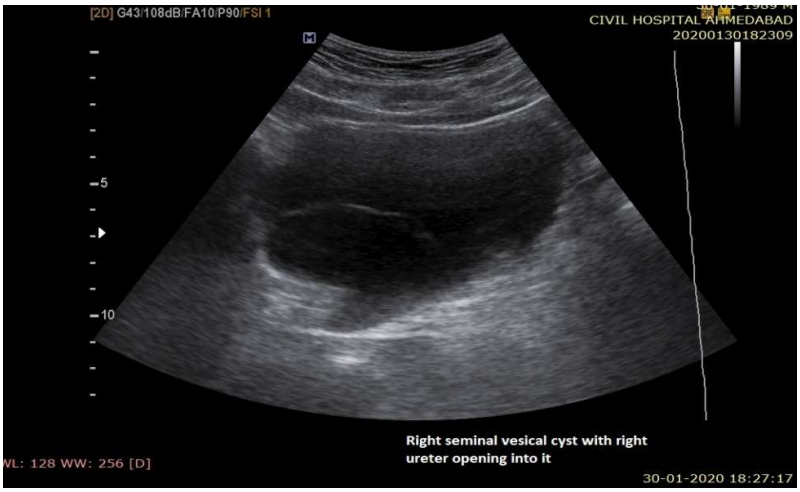


Figure 2 CT Scan of Abdomen

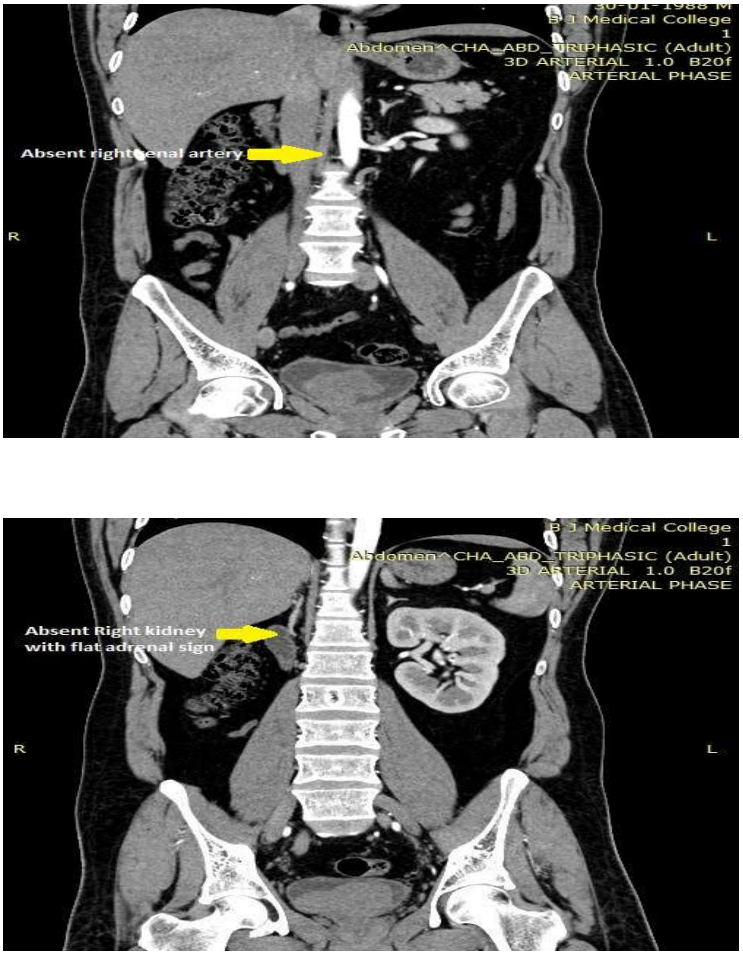
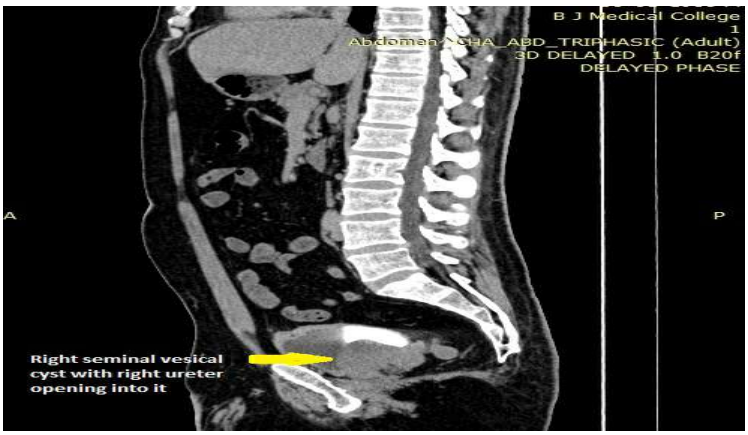
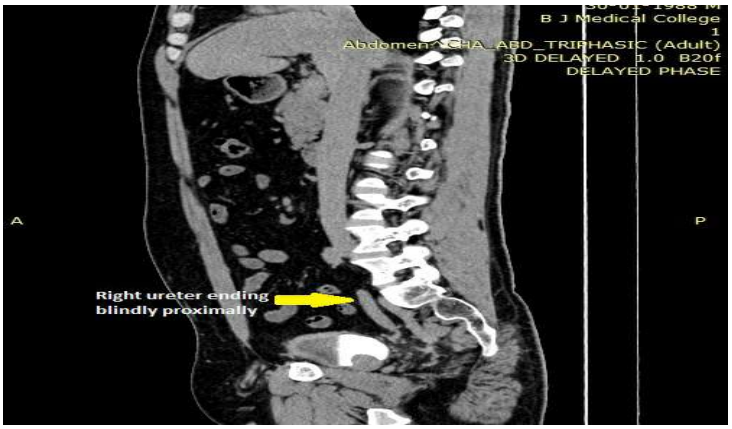


Figure 3 MRI of Abdomen





Discussion

Zinner syndrome is 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]. Zinner syndrome is rarest congenital abnormalities of the urogenital system which is usually present and diagnosed in the 2nd to 4thdecade of life[2]. 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[2]. 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. There is an association between congenital malformations of the seminal vesicle and the ipsilateral upper urinary tract because both the ureteral buds and the seminal vesicles originate from the mesonephric (Wolffian) duct[2,3]. It is a rare congenital disorder which includes other abnormalities like polycystic renal disorders, ipsilateral testicular agenesis, atresia of the vas deferens and hemivertebra. In some rare conditions, ureteric bud remnant may be associated with Zinner's syndrome. The ureter bud remnant may process initially when fetal development takes place. then, it stops due to unknown reason. Some metanephric mesenchyme development disorder may cause this situation. The different interactions between the epithelial ureteric bud and metanephric mesenchyme cause such congenital abnormalities. In many cases, ureteric bud remnant coexists and may open into seminal vesicle cyst.

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 urinary bladder and to show anechoic structures[2-5]. CT findings might include an ipsilateral renal agenesis in addition to a well defined cystic tubular lesion with bulbous anterior intra vesicle cephalic and lateral to prostate[2-6]. 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[2].

Seminal vesicle cysts should be differentiated from other cysts such as: **Prostatic utricle cyst and Mullerian duct cyst**: Prostatic utricle cyst **(**or utricular cyst**) (**PUC**)** is an area of focal dilatation that occurs within the prostatic utricle**.** Mullerian duct cyst is a cyst that arises from remnants of the Mullerian duct and is one of the midline cystic masses in the male pelvis. Prostatic utricle cysts are midline cystic masses in the male pelvis and can be very difficult or impossible to distinguish from a Mullerian duct cyst**.** Utricle cysts are most often detected in the 1st and 2nd decades of life (Mullerian duct cysts usually occur in the 3rd and 4th decades). Association of prostatic utricle cysts with a variety of genitourinary abnormalities is recognized and include: hypospadiasandcryptorchidism Mullerian duct cysts have no such associations .Prostatic utricle cysts always arise from the level of the verumontanum and are always in the midline. Mullerian duct cysts can arise anywhere along the path of Mullerian duct regression, from scrotum to utricle. Utricle cysts are variable in size but are usually smaller (commonly <10 mm) than Mullerian duct cysts and usually do not extend above the prostate gland (Mullerian duct cysts typically extend above the prostate gland).

Ejaculatory duct cyst: Ejaculatory duct cysts are a rare type of cyst of the prostate gland. They occur due to obstruction of the ejaculatory duct which in turn can either be congenital or secondary (e.g. inflammation). They are usually intraprostatic when small but may extend cephalad when large.Typically seen as round or oval lesions. They tend to be in a medial or paramedial position in the prostatic gland above the level of the verumontanum**,** extending into the prostatic base.

Treatment modality of seminal vesicle cysts 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 [7-10]. 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.

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