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Case report

Right-sided Zinner syndrome with a left side quadrupled ureter– a case report☆,☆☆,★,★★

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ABSTRACT

Zinner syndrome is a rare congenital anomaly of the genitourinary tract, consisting of ipsilateral renal agenesis, seminal vesicle cysts, and ejaculatory duct obstruction. Besides, quadruplication of the ureter is the rarest anomaly composed of proximal four ureters ending with a single distal ureter. The authors present an adult male patient with left flank pain and dysuria who was referred for abdomen CT scan. The CT scan revealed renal agenesis, seminal vesicle cysts, and obstructed ejaculatory duct, all in the right side (Zinner syndrome), and quadruplication of the ureter on the left side. The additional finding of hemivertebra was present resulting in kyphoscoliosis. Zinner syndrome and quadrupled ureter, are both rare anomalies, and the occurrence of both entities at the same patient is exceptionally rare, and have not been reported yet in the English literature. Furthermore, standard treatment protocols have to be pursued, as such patients hold a single kidney with the quadrupled ureter.

Background

Zinner syndrome is a rare congenital malformation characterized by the association of renal agenesis, seminal vesicle cysts, and obstruction of the ejaculatory duct, all of them on the same side. It was first described by Zinner in 1914 [4]. And, quadruplication of the ureter, an exceedingly rare anomaly is the existence of 4 incomplete ureters. About 15 cases have been reported up to now[6] and only 3 cases were known until early 1994 [9]. There are about 100 cases of triplication of the ureter reported in the literature [6]. Theoretically, the presence

Abbreviations: CT, Computed Tomography; MRI, Magnetic Resonance Imaging; MRKH syndrome, Mayer-Rokitansky-Kuster-Hauser syndrome; IVU, Intravenous Urography; UTI, Urinary tract infection; MCUG, Micturating cystourethrography.

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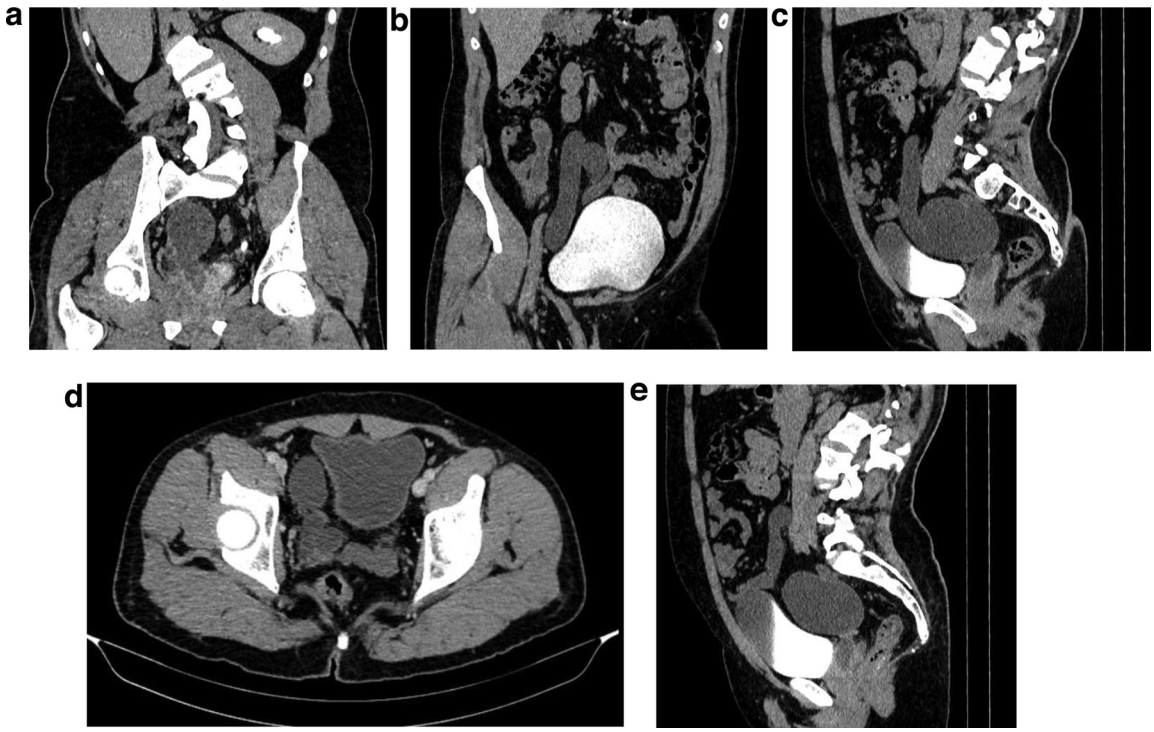

Fig. 1 – Coronal, sagittal and axial contrast-enhanced images (A) Right kidney is absent om the renal fossa. There is en enlarged seminal vesicle cyst and dilated ejaculatory duct in the right side, with abrupt narrowing at the prostate. (B) Atretic dilated right ureter (C) Atretic dilated right ureter with ectopic insertion into seminal vesicle cyst, and dilated ejaculatory duct with abrupt narrowing at the prostate. (D and E) Enlarged right side seminal vesicle with cystic formations, while in the left side normal appearing seminal vesicle.

of more than one ureteric bud may result in multiple ureters [7,14]. Patients with Zinner syndrome remain asymptomatic for a long time, and there is a strong correlation between sexual activity and the onset of symptoms [5]. On the opposite, patients with quadrifid ureter may experience recurrent UTI [6,9,10,11] Many cases of quadruplication of ureter have an association of ipsilateral ureteral cyst[6], but our case lacks such findings. Accompanied vertebral abnormalities such as hemivertebrae are also observed with genitourinary anomalies,[15] as in our case. Authors present an extremely scarce combination of congenital urinary tract malformations; Zinner syndrome on the right side and quadrupled ureter on the left.

Case presentation

A 35-year-old male patient complaining of acute left flank pain and dysuria was referred for an abdomen CT. The patient had a gross back abnormality. No previous medical, family, psycho-social history, or relevant genetic information. There was no history of previous surgery. The CT scan demonstrated agenesis of the right kidney and dilated atretic right ureter. Also, multiple seminal vesicle cysts and dilated ejaculatory duct were present on the right side. (Figs. 1A-E) The left kidney had 4 moieties from which four separate ureters were arising, where 2 central ureters were joining more proximally. All of the remaining proximal ureters were joining at the distal portion of the left proximal ureter. The distal left ureter was mildly prominent. (Figs. 2A-D) No hydronephrosis or nephrolithiasis was present. The patient also had a segmented, dorsolaterally oriented hemivertebra at D10 of the thoracic spine causing kyphoscoliosis with major convexity towards the right side. (Figs 3A-C)

After receiving the radiology report, the patient was lost to follow up.

Discussion

Zinner syndrome is a triad of Wolffian duct anomalies consisted of ipsilateral renal agenesis and seminal vesicle cysts in association with ejaculatory duct obstruction [1,5].

Up to date, there have been less than 100 cases reported in the literature regarding the Zinner syndrome. The onset of symptoms in Zinner syndrome is variable from 16 to 68 years of age[3]. As the reproductive activity increases in the 2nd to 3rd decade, the seminal vesicle cysts enlarge[4] until then most of the patients remain symptomless [5]. Zinner syndrome is considered equivalent to MRKH syndrome (Mayer-Rokitansky-Kuster-Hauser syndrome) in females [2,5]. The reason behind the developmental anomaly in Zinner syndrome is the close relationship between the Wolffian duct (responsible for a male reproductive system) and ureteric

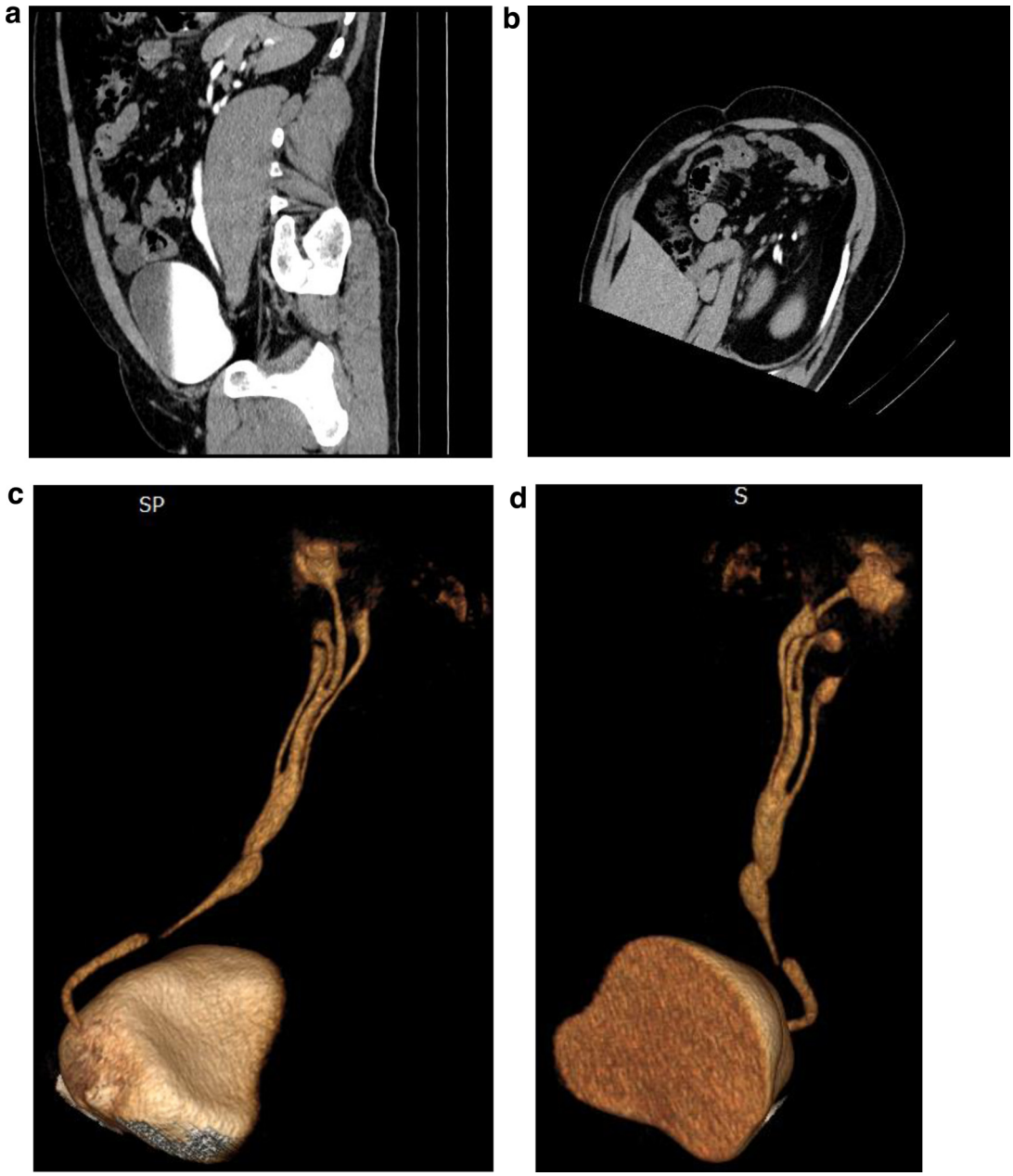

Fig. 2 – Sagittal and axial contrast-enhanced CT images (A and B) four proximal ureters opacified and distal single opacified ureter. (C and D) Spine 3D VRT showing four proximal ureters, and distal single ureter.

bud[2] The distal mesonephric duct forms the following structures: hemitrigone; urinary bladder neck; urethra up to external sphincter; seminal vesicle; vas deferens; ejaculatory ducts; epididymis; paradidymis; and an appendix of the epididymis. All of the above are under the influence of testosterone and anti-Mullerian hormone [2]. Around 4-6th week of gestational age, the metanephric blastema starts secreting growth factors that induce growth of a ureteric bud. The ureteric bud merges with a metanephric blastema resulting in the formation of a primitive kidney. Eventually, any disruption of inductive events such as mutation of metanephric blastema or disturbance of retinoic acid signaling pathway may result in agenesis of a kidney and atresia of ejaculatory ducts. Seminal vesicle cysts are then formed by the progressive accumulation of secretory products [1,2]. The commonly associated anomalies are ectopic ureter and megaureter. Usually, Zinner syndrome is diagnosed when seminal vesicle cysts enlarge[3] at least when they reach a size of 5 cm [1]. Patients frequently complain of dysuria (37%), frequency (33%), perineal pain (29%), epididymitis (27%) and pain after ejaculation (21%) (Van den Rouen et al.). However, patients may also remain symptomless or complain of perineal pain and/or infertility [1]. In a case reported by Pavan et al., a seminal vesicle cyst was visualized as a para testicular mass mimicking varicocele [1,5]. In the same year (2015), Kanavaki et al. reported a para vesical cyst as ureterocele by

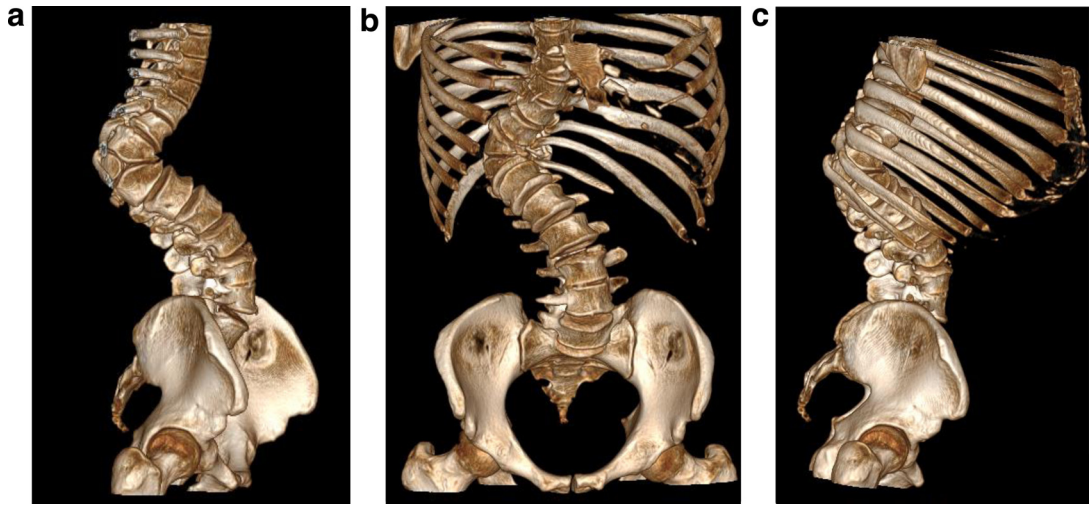

Fig. 3 – (A-C) Spine 3D VRT showing hemivertebra at the D10 with kyphoscoliosis.

ultrasound in a 4-year-old male patient, and finally after 11 years of follow-up, the diagnosis of Zinner's syndrome was established [5].

Several types of diagnostic tools can be employed to differentiate seminal vesicle cysts from other pelvic masses. Although ultrasound is a noninvasive method to assess a mesonephric duct developmental anomaly, it has a limited scope of view and is operator-dependent[1]. Anechoic structure at the site of seminal vesicle [1,2] and absent ipsilateral kidney can be seen in ultrasound [2]. CT scan is a better imaging examination, and the findings include periprostatic cystic mass located at the posterior aspect of the prostate [2]. It can accurately show renal anomalies and define pelvic anatomy [4]. Magnetic resonance imaging (MRI) is the modality of choice which can better delineate the anatomy of the male genital system and can distinguish cysts of the seminal vesicle from other pelvic masses [2]. The characteristic appearance of seminal vesicle cysts is its paramedian location. The cyst contents would return low or high T1 signal intensity (depending on protein content) and high signal intensity on T2 weighted images [2]. Usually, cysts are less than 5 cm in size [2]. There is a strong possibility of the urinary bladder and intestinal obstruction in giant seminal vesicle cysts [2,4]. Magnetic resonance imaging (MRI) is superior to computed tomography (CT) because it can better provide pelvic anatomy [3]. Vasovesiculography is another imaging tool to evaluate seminal vesicles by injecting iodinated contrast material into vas deference. Reflux of the contrast material in the ipsilateral atretic ureter can be assessed [2]. Location, shape, and compression on adjacent structures are helpful signs to distinguish seminal vesicle cysts from other cystic masses of Wolffian and Mullerian duct origins. Enlarged cysts of a seminal vesicle can protrude into the urinary bladder and compress the prostate and rectum [3]. The gold standard treatment for symptomatic patients is surgery. Open surgery is nowadays surpassed by minimally invasive techniques [5]. Kord et al. have described minimal invasive surgery as an efficient and feasible technique, more advantageous to both surgeons and patients [5].

Smith termed incomplete 3 ureters as triplication of a ureter [8]. Taking Smith's definition into account, our patient has four incomplete ureters and can be termed as a quadrifid ureter.

The duplication of a ureter is not uncommon, but triplication of a ureter is a rare anomaly [7,12,13]. Quadruplication of a ureter is an extremely rare anomaly [7,10]. Up to date, only 15 cases have been reported in English literature and 11 of them had surgical treatment by ureteral reconstruction, excision or implantation of ureter, nephrectomy, or heminephrectomy [6].

The primordian of the collecting system is a ureteric bud that develops from the Wolffian duct in the 5th week of gestation [8]. The formation of multiple ureters is thought to be related to the early division of the ureteric buds. Then the metanephric tissue divides into multiple parts and each renal pelvis with its ureter [7,8]. The development of more than one ureteric bud signifies duplication or triplication of a ureter [13,14].

Most of the patients with ureteric quadruplication have recurrent UTI [6,9,10,11]. In our case, the patient was complaining of left flank pain and dysuria for a week.

The associated anomalies include ureteral cyst and vesicoureteral reflux. 9, but our case showed no findings of a ureteral cyst.

The preoperative diagnosis of a quadrifid ureter can be made by employing IVU, ultrasound, MCUG, MAG3, or MR urography. Intraoperative diagnosis of a quadrifid ureter can be achieved with retrograde pyelography, laparotomy, or laparoscopy [6].

Nephrectomy, heminephrectomy, and cutaneous ureterostomy are the utilized surgical techniques in those patients with recurrent UTI [11].

Developmental vertebral anomalies include hemivertebrae, wedge vertebrae, block vertebrae, and butterfly vertebrae, to name a few of them. Lack of formation of one-half of a vertebral body is termed hemivertebrae. It can occur sporadically, or in association with various other anomalies including anomalies of the genitourinary system, as in our case. Hemivertebra has various forms; (1) fully segmental, where it

is not attached to either vertebra above or below, as in our case, (2) semisegmental, where a half segment is attached to the vertebra above or below without the presence of intervertebral disc, (3) nonsegmental, where it is attached to either vertebra above or below, and (4) incarcerated type, where there is an attachment by pedicles to levels above or below. It has also orientational forms such as dorsal, lateral, or ventral hemivertebra. Our case appears as a dorsolateral hemivertebra with resultant kyphoscoliosis.

A wedge-shaped vertebra may mimic wedge shaped fracture. It is a congenital anomaly where there is a reduction in the height of the anterior aspect of a vertebral body. Block vertebra is a fusion of 2 or more vertebrae, either ventral (body), dorsal (arch), or both. Klippel-Feil syndrome is a partial or complete fusion of 2 or more cervical vertebrae. A butterfly vertebra can result from a non-fusion of the lateral halves of the vertebral bodies [15].

Zinner syndrome and quadruplication of a ureter are both rare anomalies, and the combination of both anomalies in a single patient is an exceptionally rare finding. Further studies may be needed to pursue a right therapeutic approach especially in patients who only hold a single kidney with a quadrupled ureter.

Ethics approval and consent to participate

The manuscript has got ethical review exemption from Ethical Review Committee (ERC) of the authors' institution (French medical institute for Mothers and Children- {FMIC}) as case reports are exempted from review according to the institutional ethical review committee's policy.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study (as this is a case report).

Authors' contributions

All of the authors have participated sufficiently in the submission and take public responsibility for its content. NF: writing and editing the manuscript, selecting the images, and corresponding with the journal. MNN: Revising the manuscript and selecting the case. All of the authors have read and approved the final manuscript.

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