

Zinner Syndrome: A Rare Case of the Mesonephricduct Anomaly

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Abstract

The present article reports 18-year-old boy came to the urology clinic with perineal discomfort and few episodes of incomplete emptying of the bowel for two months. He did not claim any symptoms related to his voiding or ejaculation. The abdominal and external genitalia examination was regular. He had average prostate size on digital rectal examination with a palpable painless cystic mass just above the prostate gland. Zinner's syndrome should be a differential diagnosis in young patients with urinary symptoms and unilateral renal agenesis. A detailed review of the relevant literature is also presented.

Keywords: Zinner's syndrome; congenital malformation; seminal vesicle cyst; renal agenesis; infertility.

Introduction

Zinner's syndrome is one of the rarest congenital malformations present with cysts in the seminal vesicle, ejaculatory duct obstruction, and ipsilateral renal agenesis [1]. Its origins in the development abnormality of the Wolffian duct at embryogenesis. Some of the patients with Zinner's syndrome may remain asymptomatic and discovered incidentally, while others present with clinical signs of bladder outlet obstruction, nonspecific pelvic pain, and symptoms related to ejaculatory dysfunction [2]. Herein, we present a case of Zinner's syndrome in which the patient presents with perineal pain and discomfort with a few episodes of incomplete defecation as an initial complaint.

Case presentation

An 18-year-old boy came to the urology clinic with perineal discomfort and few episodes of incomplete emptying of the bowel for two months. At the same time, he did not claim any symptoms related to his voiding or ejaculation. The abdominal and external genitalia examination was regular. He had average prostate size on digital rectal examination with a palpable painless cystic mass just above the prostate gland. The initial laboratory investigations did not reveal any abnormal values, including renal function tests and hormone analysis (LH, FSH, and testosterone). The ultrasound kidney, ureter, and bladder (KUB) showed the right side renal agenesis and homolateral seminal vesicle cyst (Figure 1). Subsequently, the urinary tract's magnetic resonance imaging (MRI) was requested, which diagnosed Zinner's syndrome by visualizing the seminal

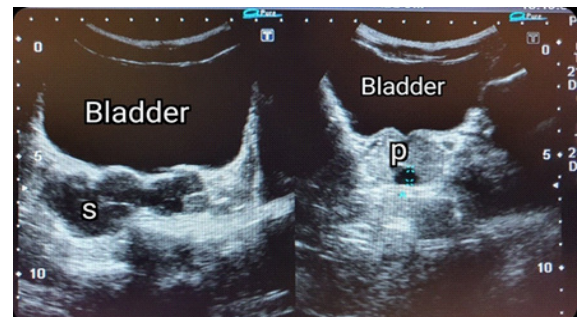


Figure 1: Ultrasound pelvis showing the multiple cysts within the right seminal vesicle(S) and dilated distal part of seminal vesicle within prostate gland (P).

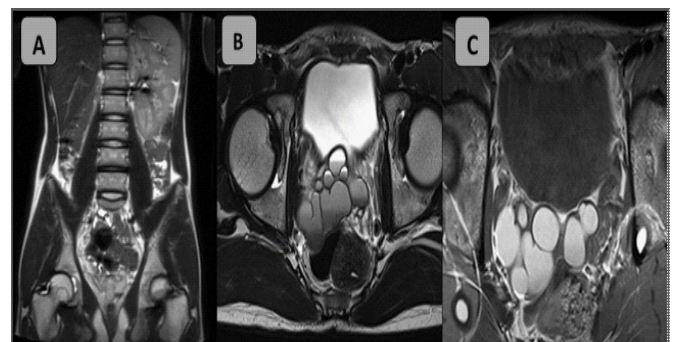


Figure 2: (A) Coronal image of MRI abdomen showing absent right kidney with enlarged normal left side kidney. (B & C) Pelvic MRI. T1 and T2 – weighted axial images show the right side dilated cysts within the seminal vesicle, and it shows hyperintensity in T1 and hypointensity in T2-weighted images.

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vesicle cyst on T1 and T2 weighted images as hypo intensity and hyper intensity signals, respectively (**Figure 2**). Furthermore, his sperm parameters were normal in limit and excluded the risk of future infertility. We offered simple nonopioid analgesics for the perineal pain and laxatives for the defecation issues. We did not do any invasive treatment for him as he is asymptomatic after initial treatment. Therefore, he is under surveillance at our urology clinic for more than one year.

Discussion

Zinner's syndrome occurs due to abnormal growth of the Wolffian duct during embryogenesis, and it is marked by ejaculatory duct obstruction, cysts in seminal vesicles, and same-side renal agenesis [3]. In history, the seminal vesicle cysts were identified first by Smith in 1872, and later the association between unilateral renal agenesis and seminal vesicle cyst was first described by Zinner in 1914 [2]. Since that, more than 100 case reports in Zinner's syndrome reported in the English literature and this is the first reported case in Sri Lanka in this entity [4]. The ureteric bud originates from the distal part of the mesonephric duct and ascends cranially to meet the metanephric blastema, which will form the future kidney. Maldevelopment of the distal mesonephric duct occurs due to various insults during the first trimester that leads to ejaculatory duct atresia and abnormality of ureteral budding. Atresia of the ejaculatory duct causes obstruction and cystic dilatation of seminal vesicles and anomaly in the ureteral budding results in renal agenesis [5]. Patients with this congenital anomaly are usually asymptomatic until their second to the third decade of life [6]. The symptoms develop due to bladder irritation, bladder outlet obstruction, cyst distension, and obstruction of the ejaculatory duct. Therefore, the Patients may present with unspecific and various clinical manifestations, including voiding symptoms (frequency, dysuria, urgency, poor flow), haematuria, pelvic pain, perineal or scrotal pain or discomfort, urinary tract infection, painful ejaculation, and haemospermia [5].

The diagnostic evaluation of a Zinner's syndrome includes biochemical investigations, radiological imaging, and cystoscopic examination. Urinalysis and culture, blood analysis, renal function tests, and hormone profile (FSH, LH, Testosterone) is proper initial biochemical investigations. Transrectal ultrasonography is the most widely used tool for identifying and initial evaluating seminal vesicle cysts, and it reveals anechoic cystic pelvic lesions with a thick, irregular wall and calcifications [7]. Besides, the computed tomogram also can demonstrate the cysts in the seminal vesicle and renal agenesis, but it is not adequate to make the final diagnosis. Therefore, magnetic resonance imaging is the definitive diagnostic imaging of Zinner's syndrome without the need for additional invasive investigations. Typically, the cysts in seminal vesicles appear as hyperintensity lesions on T2- weighted images and hypointensity in T1- weighted images. Besides, an MRI scan reveals excellent soft-tissue anatomy between the cysts and surroundings in the pelvis, which is helpful for surgical management [3]. Urethrocytoscopy may show trigonal abnormality or bulge inside the bladder due to external compression [8].

The management of Zinner's syndrome should be planned

according to the clinical presentation. The Conservative approach is helpful in mild symptomatic or asymptomatic cases with normal biochemical parameters [6]. Aspiration of the seminal vesicles and combined instillation of substances such as alcohol or minocycline and antibiotics are also tried in mild symptomatic group 3. Surgical treatment options should restrict to symptomatic cases or patients who failed conservative measures. Surgical treatment options can be a transurethral resection of the ejaculatory duct (TURED) or seminal vesiculectomy. TURED procedure includes the resection of the prostate at the level of verumontanum until the opening of the ejaculatory ducts. A study reported a remarkable improvement of semen volume, pH, and sperm count, increasing carnitine and fructose concentration after TURED in seven patients. Therefore, they recommended that TURED surgery be tried first to resumption natural fertility before trying other infertility treatments [9]. Open excision of the cyst was the most effective procedure for symptomatic patients until recently, and it was replaced by laparoscopic and robotic surgeries in the current era. Minimally invasive surgeries ensure retrovesical anatomy during surgery with minimal blood loss and minimal postoperative morbidity. Unfortunately, some of the patients with infertility fail to recover from poor sperm parameters despite all these efforts, and assisted reproduction techniques become the only hope for them [10].

Conclusion

Zinner's syndrome should be a differential diagnosis in young patients with urinary symptoms and unilateral renal agenesis. Even though transrectal ultrasonography is used as an initial screening tool, the MRI pelvis makes the final diagnosis, and surgical treatment options resolve the complaints in symptomatic patients except for the poor sperm parameters.

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