Zinner’s Syndrome: A Rare Case Series

Hiranya Deka, Rohit Kumar Singh and Appu Thomas*
Department of Urology, Amrita Institute of Medical Science, India

Abstract

Zinner’s syndrome is a very rare congenital condition characterized by seminal vesicle cyst, with upper tract anomaly—common being ipsilateral renal agenesis or renal dysplasia or ipsilateral congenital absence of vas deference is also present and obstruction of the ejaculatory duct. This condition would be a diagnostic challenge and its management at different age groups is also challenging as the indication for treatment will differ with age. Here we present a case series of two case of different age group with their different presentation and management strategies.

Keywords: Zinner’s syndrome; Infertility; Renal agenesis, Seminal vesicle cyst; Ejaculatory duct obstruction

Introduction

Zinner’s syndrome is a very rare congenital condition characterized by an ipsilateral absent kidney, seminal vesicle cyst, and ejaculatory duct obstruction from a developmental anomaly of the Wolffian duct. Kidney and seminal vesicle have a common embryological origin and anomaly of Wolffian duct can result in genital tract maldevelopment and ureteric bud abnormality can result in non-attachment to metanephric blastema. This is the embryological basis of Zinner’s syndrome.

Case Series

Case 1

A 26 years old short-statured young male presented with recurrent left lower abdominal pain. He underwent Ventricular Septal Defect (VSD) and Aortic Regurgitation (AR) repair in infancy due to congenital valvular heart disease. He also had recurrent left testicular pain since puberty. He got married 2 years back, had no erectile dysfunction but complaints of poor volume ejaculate and has not been able to impregnate. Ultrasonography abdomen showed a normal right kidney with the absent left kidney and there was a cystic lesion behind and to the left of the urinary bladder. Contrast-Enhanced Computed Tomography (CECT) and Magnetic Resonance Imaging (MRI) confirmed the ultrasonographic findings as a left seminal vesicle cyst with an absent left kidney (Figure 1, 2). Semen analysis showed one milliliter ejaculate with severe oligoasthenospermia. The cyst was laparoscopically deroofed which relieved the pain of the patient. He is planned for TESA as a treatment plan for his infertility.

Case 2

A 16 years old young male presented with intermittent lower abdominal pain. Ultrasonography showed cystic lesion behind the bladder and ipsilateral non visualized kidney and MRI did which confirmed the diagnosis of seminal vesicle cyst with ipsilateral renal agenesis (Figure 3). He was...
managed conservatively with medication without any intervention. His fertility would be assessed at an appropriate time.

**Discussion**

Zinner’s syndrome was first described by Zinner in 1994 and very few cases were reported in world literature [1]. Incidence is 1 in 4000 newborns. The syndrome can be explained as a wolffian duct (which develops the male reproductive system) anomaly and absence or mutation of ureteric bud which failed to fuse with metanephric blastema leading to renal agenesis [2]. Usually, this condition is asymptomatic but for symptomatic patients, treatment may be offered [3]. Many of these patients have oligoasthenospermia which results in infertility and can be planned for TESA to father their child. Genetic co-relation has to be clarified before starting infertility treatment.

**Conclusion**

The triad of Zinner’s syndrome i.e. ipsilateral renal agenesis, seminal vesicle cyst and ejaculatory duct obstruction is a very rare condition. A high index of suspicion is required to unearth this condition more frequently. Symptomatic management can be offered with a minimally invasive procedure. Infertility is a more serious sequela that also needs to be addressed along with symptomatic management.

**References**