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Case report

Zinner's Syndrome as a Rare Case Report

Hrishikesh Tovar[1], Sneha Karhade[2], Anant Beedkar[3], Sarojini Jadhav[4]

[1]Junior Resident, Department of General Surgery, Govt. Medical College, Aurangabad.
[2]Senior Resident, Department of General Surgery, Govt. Medical College, Aurangabad.
[3]Acadamic Professor, Department of General Surgery, Govt. Medical College, Aurangabad.
[4]Professor and HOD, Department of General Surgery, Govt. Medical College, Aurangabad.

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Corresponding author: Dr. Hrishikesh Kiran Tovar
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Abstract

Introduction: Zinner's syndrome is a triad of mesonephric (wolffian) duct anomalies comprising of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. Maldevelopment of distal part of the mesonephric duct results in atresia of ejaculatory duct (leading to obstruction and dilatation of seminal vesicle), while abnormality in ureteral bud leads to renal agenesis.

Method: Here we present a case of 17 years old male child with complain of pain in abdomen on right side. USG & CECT[A+P] was done which confirmed above diagnosis. He was managed by exploratory laparotomy with right abdominal testis and cyst excision and was sent for HPE. Post operative period was eventful. Histopathology report was s/o cryptorchid testis.

Results and Conclusion: Seminal vesicle cyst combined with ipsilateral renal agenesis is a rare anomaly in the development of urogenital system. This usually occurs in male in 2nd to 4th decade of life. The usual presentation is pain in abdomen, perineum and scrotal region. The diagnostic workup consists of Transrectal USG, CECT, MRI pelvis. The definite treatment is open surgery.

Keywords: Seminal Vesicle, Cyst, Exploratory Laparotomy, Cryptorchid Testis, Zinner's Syndrome.

Introduction

Zinner syndrome is a rare disease, affecting males. It is secondary to an abnormal evolution of the mesonephric (Wolffian) duct during embryogenesis. The syndrome is characterized by atresia of the ejaculatory duct, leading to its dilation and/or seminal vesicle cysts, associated with ipsilateral renal agenesis. [1],[2] Mainly due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct. [3],[4] Most of the patients with Zinner syndrome are asymptomatic. Clinically it is generally manifested by ejaculatory disturbances, local pain, and infertility, but it can display a wide range of presentations. [5],[6] Patients with Zinner syndrome commonly present with infertility, dysuria, urgency, prostatism and painful ejaculation. The syndrome may often present with voiding symptoms after during the period of utmost sexual or reproductive activity i.e., mostly in second

and third decade of life.[7] The diagnosis may be set by ultrasonography or CT scan, but MRI gives a complete image of the local anatomy.[8] Selection of treatment options mainly depends on the presentation of symptoms. The treatment strategy mainly focuses on seminal vesicle cyst. Patients are mostly managed conservatively. The treatment of choice for seminal vesicle cysts is surgical resection and other methods such as transurethral puncture, aspiration, transurethral resection of seminal colliculus and vas deferens.[7] Zinner syndrome may lead to infectious complications, such as seminal vesicle abscess, or it may be associated with other developmental abnormalities or tumors. [1] It was first reported by Zinner in 1914. [9] Till now only few hundreds of cases reported in the literature. [3] Hereby we like to present about the rare developmental anomaly involving the Mullerian ducts encountered in our hospital.

Case Scenario:

A 17 years old male child with complain of pain in abdomen on right side for 1 month. There was no history of vomiting, fever, blunt trauma, hematuria. Clinically per abdomen examination revealed mild tenderness in right iliac fossa with no palpable lump. Genital examination revealed empty right scrotal cavity. USG(A+P) was s/o large hypoechoic lesion of 15x10x8 cm in pelvis and absent right kidney and USG scrotum s/o absent right testis. CECT[A+P] revealed large cystic lesion of size 15x10x8 cm in pelvis indenting base of bladder with absent right kidney and undescended right testis. Semen analysis revealed marked Oligospermia with spermosthenia.

He was managed by exploratory laparotomy with right abdominal testis and cyst excision and was sent for HPE. Large retroperitoneal cyst of size 20x10x10 cm in retro vesical space, occupying right renal fossa completely and right ectopic testis of size 2.5x1x1 cm adherent to cyst wall. Post operative period was eventful. s /o cryptorchid testis.

Histopathology Examination:

Histopathology report showed seminal cyst and cryptorchid testis respectively.

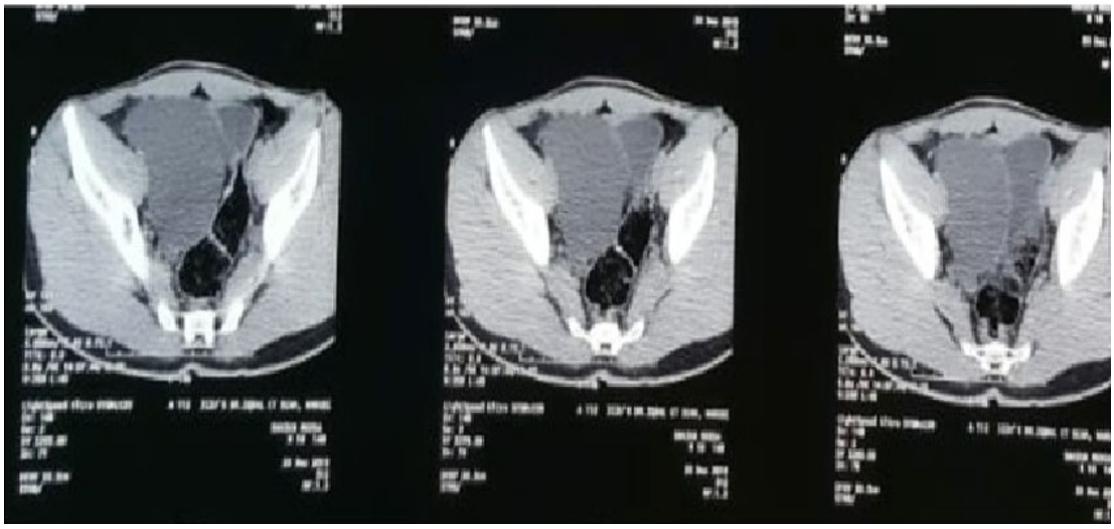

Figure 1: Pre operative CT

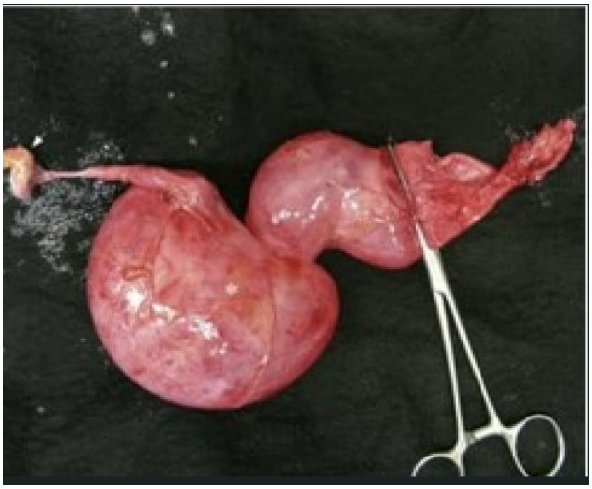

Figure 2: Excised seminal vesicle cyst with right testis

Discussion

In an ultrasonographic screening performed on 280,000 children over 2 and a half years in Taiwan in 1990, 13 cases of Zinner syndrome were identified, thus estimating a frequency of

0.00464% [10]. In a systematic review of the publications on Zinner syndrome between 1999 and 2020, a Chinese group of researchers counted 214 cases. Its pathogenetic mechanism consists in a failure of the mesonephric duct to develop, between the 4th and the 13th week of intrauterine life, in both directions: upwards, where there is no fusion between the ureteric bud and the metanephric blastema, which is hence not induced to form the kidney, and downwards, where the permeability of the ejaculatory duct is affected [5].

In the adult population the most frequent clinical presentation is that of our patient, i.e., urinary symptoms like dysuria, painful micturition, urgency, incontinence, hematuria. The next group of symptoms as frequency of appearance is local pain: perineal, lower abdominal, pelvic, and scrotal pain. Disturbances in the reproductive function are also described: infertility, painful ejaculation, decrease of the volume of the semen. Hematospermia may be the presenting symptom [11]. Similar complaints were seen in our case.

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. 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] We also completed CT abdomen-pelvis and got to know the complete image of the local anatomy and extension of lesions.

According to the most complete study, the pooled analysis of the Chinese group on 214 cases, most of the patients (65.8%) underwent surgical interventions (seminal vesiculectomy, unroofing of cysts etc.). It is worth noting that most of the interventions were minimally invasive, using the laparoscopic or even robotic approach. The outcome of most of the patients is favourable. Out of the 214 patients in the pooled database of the Chinese researchers, 52 patients had complications (mainly infections, even sepsis) and comorbidities. [1]

According to Florim et al and Célia Sousa1 (2019) had the same view that ., the management of this syndrome should be clinically oriented and it can range from follow-up in asymptomatic and minimally symptomatic cases, and that is what was preferred by our patient (otherwise, antibiotics, transurethral aspiration of the

seminal vesicle cyst, or transurethral aspiration combined with substance installation (e.g., alcohol and minocycline) is proposed as the conservative treatment), up to invasive treatment that should be considered only in symptomatic patients or patients who failed conservative measurements and then usually consists of transurethral resection or balloon dilatation of the ejaculatory duct and open or laparoscopic vesiculectomy. [12],[13]

According to the literature reviewed and the clinical and radiological presentation of our patient we performed exploratory laparotomy with right abdominal testis and cyst excision.

Conclusion

Seminal vesicle cyst combined with ipsilateral renal agenesis is a rare anomaly in the development of urogenital system. This usually occurs in male in 2nd to 4th decade of life. The usual presentation is pain in abdomen, perineum and scrotal region. The diagnostic workup consists of Transrectal USG, CECT, MRI pelvis. The definite treatment is open surgery.

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