



Case Report – Zinner Syndrome- A Rare Case Report

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Introduction

Zinner syndrome is a very rare wolffian duct abnormality characterized by congenital unilateral renal agenesis that is absence of one kidney, seminal vesicle cyst and ipsilateral ejaculatory duct obstruction . The condition was first described by zinner in 1914 [1,4] .It usually presents in third or fourth decade of life with the complaint of infertility[2] . But here we present a case of 45 year old male who presented with pyelonephritis and ist time detected Zinner Syndrome .

Zinner Syndrome is a very rare congenital abnormality believed to be the male counterpart of Mayer-Rokitansky-Kuster-Hausler Syndrome which occurs in females. The usual appearance of this abnormality dates to the fetal life between fourth and thirteenth week of gestation[1,2,3] . Majority[NEARLY 66%] of the patients present with unilateral renal agenesis and seminal vesicle cyst incidence is 0.005% .It consists of a triad of unilateral renal agenesis, seminal vesicle cyst and ejaculatory duct obstruction .The cystic mass expands with installment of sexual life , with a right/left side incidence of 2:1 . The diagnosis is established usually in the fourth decade of life .[1,3]

Case Report

A 45 year old male with no underlying comorbidity presented with complaints of fever with chills , dysuria

and right flank pain for two weeks with no other significant past history. The patient has 1 child who is eighteen years old

On examination the external genitalia were normal but per abdominal examination revealed a large ballotable mass in right lumbar area which was non tender.

On routine lab investigations his total leucocyte count was increased to 18600 with azotemia [S.Cr -1.66] . Rest all other baseline investigations were normal . On ultrasonography of abdomen we found that the patient has a solitary kidney with size of 12.9 mm / 5.8 mm . The patient was subjected to NCCT-KUB which revealed bulky right kidney with mild perinephric fat stranding. There was a cystic lesion in left seminal vesicle measuring 25/ 26 mm in size s/o seminal vesicle cyst with features suggestive of Zinner Syndrome.

The patient was managed conservatively on intravenous antibiotics [MEROPENEM and levofloxacin] till he improved.

Discussion

The zinner syndrome is defined by a triad of renal agenesis, seminal vesicle cyst and ipsilateral ejaculatory duct obstruction. It is a rare congenital condition with less than 200 cases reported worldwide. [1]

Most patients with this group of mesonephric duct anomalies are asymptomatic until the third or fourth

decade of life and often manifest during the period of high sexual or reproductive activity. The seminal vesicle cyst is asymptomatic when less than 5 cm and discovered incidentally on DRE.[1,2]

Most patients are diagnosed by imaging and differentiation from other pelvic cysts requires a multimodality approach .[5]

The patients present with dysuria, pelvic pain, painful ejaculation , chronic recurrent epididymitis, prostatitis and occasionally infertility.[2]

However presentation as acute pyelonephritis in literature seldom exists.

Conclusion Zinner Syndrome is a very rare urological congenital condition with less than 200 cases around the globe . It must be suspected in males with pelvic pain syndrome, pelvic mass , unilateral renal agenesis or infertility [MOST COMMON PRESENTATION]. [1,3]

References

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