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ZINNER SYNDROME – A CASE REPORT OF A DEVELOPMENTAL ANOMALY WITH PRESENTATION IN AN ADULT

Urology

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ABSTRACT

Zinner syndrome is triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction which is due to abnormal development of wolffian duct .Incidence rate falls between 0.00035% and 0.0046%. It is Associated with infertility in 45% of cases. That's why we need to focus on early diagnosis and treatment and shift our focus from symptomatic relief to fertility preservation. We present a case of 22 year old patient with zinner syndrome.

KEYWORDS

Zinner syndrome, infertility

INTRODUCTION

Zinner syndrome , which is a rare disease results from a abnormal development of mesonephric (wolffian duct) . Incidence rate falls between 0.00035% and 0.0046%. This disease occurs when the insult to wolffian duct is between 4th to 13th week of gestation .It is characterised by triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction . Most patients develop symptoms in 3rd to 4th decade of life . But some can lead to symptoms in early age also . Patient can present with wide range of symptoms related to LUTS , lower abdominal pain , pelvic pain , infertility . Diagnosis is made by USG , CT urography / MRI .

CASE REPORT

A 22 year old patient came with complaints of increased frequency of micturition and lower abdominal pain. On physical examination slight tenderness was present in hypogastrium and external genitalia was found to be normal. On rectal examination anal tone was normal, prostate was flat and a firm ,non tender swelling was palpable whose upper boundary can't be reached. Urine analysis and CBC was normal.

Usg abdomen revealed an empty right renal fossa, a normal left kidney and a cystic swelling of size 104*60 mm posterior to the bladder. On CT scan – right kidney was absent,? Rt renal agenesis, an enlarged left kidney of 13.4*5.6 cm , a well defined hypodense area of 12.4 *7.6 cm in rectovesical pouch ? Seminal vesicle cyst as shown in figure 1.

Semen analysis pre operative volume of 0.2 ml, pH of 8 , total sperm count 0 (azoospermia) .

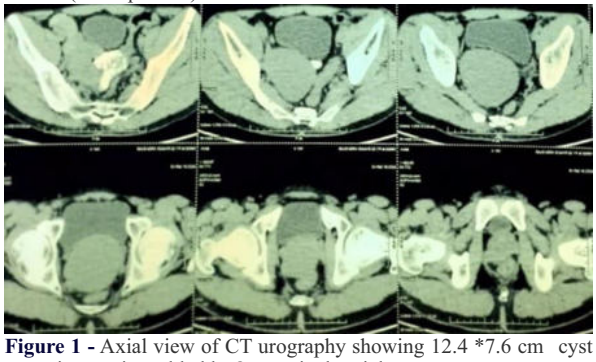

Figure 1 - Axial view of CT urography showing 12.4 *7.6 cm cyst posterior to urinary bladder ? rt seminal vesicle cyst .

MANAGEMENT

Cyst excision was done through open approach . The cyst was excised using transperitoneal approach with a vertical midline incision as shown in figure 2.

On HPE it confirmed seminal vesicle cyst.

Repeat semen analysis after 6 months had volume of 2ml, colour milky white, Ph of 8 and concentration 1 million /cumm sperms (normal 15 million / cumm) total sperm count 4 million (normal > 39 million), total and progressive motility 0, normal form 20 %, abnormal forms 80% .

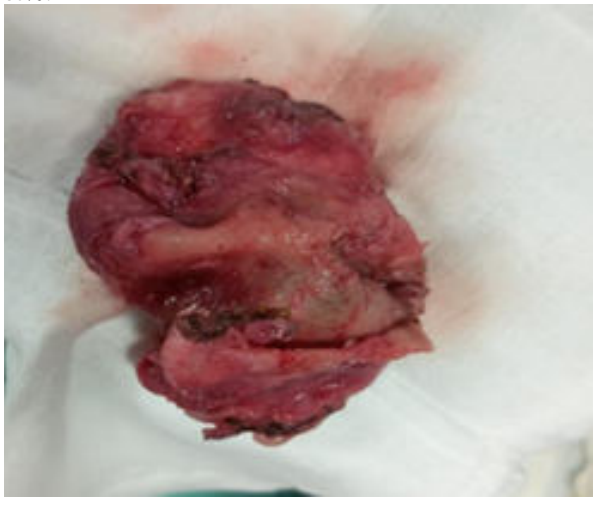

Figure 2 - Excised specimen showing Rt seminal vesicle cyst

DISCUSSION

Zinner syndrome is a very rare developmental anomaly with very few cases reported. Seminal vesicle cyst was 1st described by Smith in 1872. Association of renal unilateral agenesis and homolateral seminal vesicle cyst was 1st studied in 1914 by Zinner. Zinner syndrome leads to atresia of the ejaculatory duct, the insufficient drainage of the gonads results in cystic dilatation of the seminal vesicle.

Cysts that are generally of size more than 5 cm are symptomatic. Diagnosis is made by high index of suspicion when a male patient presents symptoms of lower urinary tract associated with renal unilateral agenesis. Another important aspect is association with male infertility (45% of cases), so semen analysis should always be done in the presence of a Zinner's syndrome.

Infertility may be caused by ejaculatory duct obstruction, which in turn results in a very low ejaculatory volume of less than 1 ml, azoospermia, low fructose and alkaline pH in the seminal plasma.

If CT scan is not able to diffrentiate seminal vesicle cyst than MRI can be helpful. Seminal vesicle cysts appear hyperintense on T2-weighted images and hypointense on T1-wieghted image. Management is follow-up in asymptomatic and minimally symptomatic cases . Antibiotics with or without transurethral aspiration of the seminal vesicle cyst or transurethral aspiration are proposed as conservative treatment. Surgical treatment is limited to

symptomatic cases or who have failed conservative measures and usually includes open, laparoscopic or robotic vesiculectomy.

In this case the semen parameters improved but not enough and oligoasthenoteratospermia persisted after the surgery. This is because of late presentation and diagnosis. In Zinner syndrome only one ejaculatory duct is obstructed then question arises that why there is infertility. Main reason is due to antisperm antibody production. By early diagnosis and treatment this infertility can be avoided .

CONCLUSION

Seminal vesicle cysts combined with ipsilateral renal agenesis that is zinner syndrome is a rare anomaly. The diagnostic work-up includes various imagings like transrectal and abdominal ultrasonography, CT scan and MRI. By diagnosing the condition early and excision before excess enlargement of cyst we can more commonly use minimal invasive technique and can also avoid infertility.

Now the focus of surgical treatment on pain relief is changing towards fertility preservation by early diagnosis and treatment which can prevent deleterious effect on contralateral testis .

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