E-ISSN 2250-0944

ISSN 2250-1150
doi: 10.48047/ijprt/15.02.313

Research Article

Case Series

Zinner Syndrome: A Series Depicting Rare Anomaly of the Male Genitourinary Tract.

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Running Title*: Zinner Syndrome: Rare Male Genital Tract Abnormality.*

ABSTRACT

Zinner syndrome is a developmental amalgamation, characterized by the presence of three congenital anomalies: unilateral seminal vesicle cyst, ejaculatory duct blockage and unilateral renal agenesis. The condition primarily arises due to developmental malformations of the mesonephric (Wolffian) duct and typically presents in the second to fourth decade of life.

In the index series, three male patients aged between 27 and 34 years presented with varied urological symptoms including dysuria, lower abdominal pain, painful ejaculation and infertility. All cases underwent thorough clinical evaluation, laboratory investigations and diagnostic imaging: Ultrasound, CT urography, and MRI—to establish the diagnosis. Findings consistently revealed unilateral renal agenesis with ipsilateral seminal vesicle cysts. One patient was treated conservatively, whereas the other two underwent laparoscopic vesiculectomy.

This paper aims to present the condition focusing on clinical presentation, radiological findings and surgical management through three distinct cases, highlighting the critical role of imaging, specifically through MRI for providing superior anatomical detail. Surgical intervention with minimally invasive approach yielded favorable outcomes, with improved quality of life and reproductive potential.

Keywords: Zinner syndrome, Renal agenesis, Seminal vesicle cyst, Ejaculatory duct obstruction, Infertility.

Introduction

Zinner first described this condition in 1914 [1]; it is characterized by unilateral renal agenesis, ipsilateral obstruction of ejaculatory duct and ipsilateral seminal vesicle cyst. The condition typically presents during the second to fourth decades of life and the affected individual could either be symptom-free or present with symptoms. A thorough knowledge of the condition, proper physical examination and imaging make it simple to establish the diagnosis [2].

The clinical condition basically stems from blocked ejaculatory duct leading to a buildup of Seminal fluid that eventually causes enlargement and appearance of cyst in the ipsilateral seminal vesicle. The patient could present with perineal discomfort, dysuria, pollakiuria, epididymitis and pain during ejaculation [3,4]. It has been revealed that up to 45 percent of men with Zinner syndrome may be infertile; cases of malignant change in the seminal vesical cysts have also been described [5].

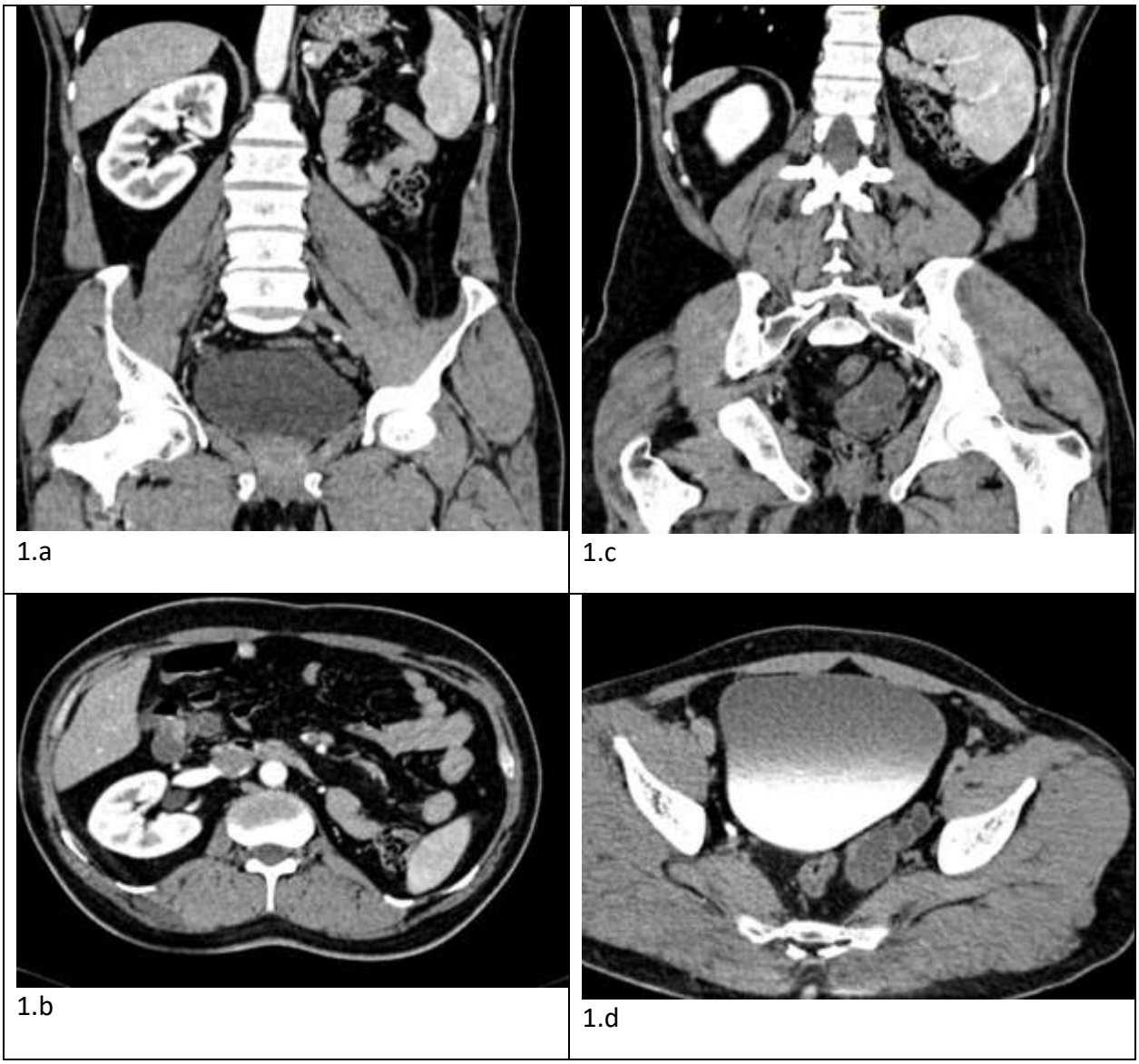
This paper highlight the clinical signs and the key imaging features of Zinner syndrome, in the contemporary era with availability of modern imaging tools and also briefly touches the management of the condition.

Case Series

Case 1:

A 34-year-old male with no significant past history presented to our hospital emergency department with complaints of lower abdominal pain, pelvic fullness and dysuria. On physical examination, there was mild suprapubic tenderness. His Complete Blood Count (CBC), Blood urea, Creatinine and Urinalysis were within normal limits. Transabdominal Ultrasound (USG) revealed normal right kidney, absence of the left kidney with no evidence of ectopia, and a cystic lesion on the left side located behind the urinary bladder.

A Contrast Enhanced Computed Tomography (CECT) was performed, which revealed left renal agenesis with intestinal loops occupying the left renal fossa and a cyst in the left seminal vesicle (Fig.1). Based on these observations, a diagnosis of Zinner syndrome was made and the patient was treated with a conservative approach.



1.a 1.c

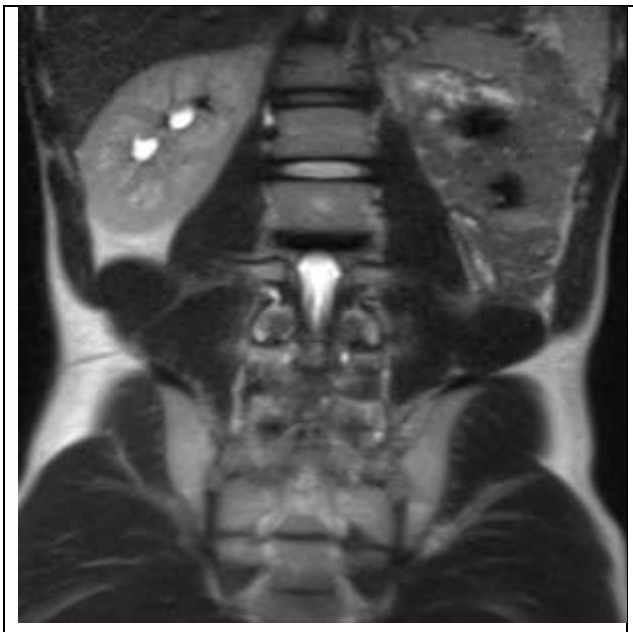
1.b 1.d

Fig 1. (a,b) Coronal and axial CT scans show an empty left renal fossa, indicating the absence of the left kidney; and **(c,d)** coronal and axial CT images revealing non-enhancing lobulated left retro-vesical cystic structure diagnostic of seminal vesicle cyst

Case 2:

A 30-year-old male presented to our hospital Out Patient Department (OPD) with complaints of pelvic pain, painful ejaculation and infertility for 1-year. There was no significant past illnesses, or any history of previous trauma or surgery. On physical examination, normal secondary sexual characteristics and normal external genitalia were present. Semen analysis revealed

normospermia. He was reffered to the Radiology department for infertility workup. On UGS, there was a solitary right kidney and cystic tubular structure in pelvis, posterior to the bladder. Abdominal MRI was done which revealed agenesis of left kidney and T1- hypointense, T2- hyper-intense lobulated cystic structure in the pelvis consistent with left seminal vesicle cyst (Fig.2). Based on the clinical, imaging and laboratory findings a diagnosed of Zinner syndrome was made The patient later underwent Laparoscopic vesiculectomy.



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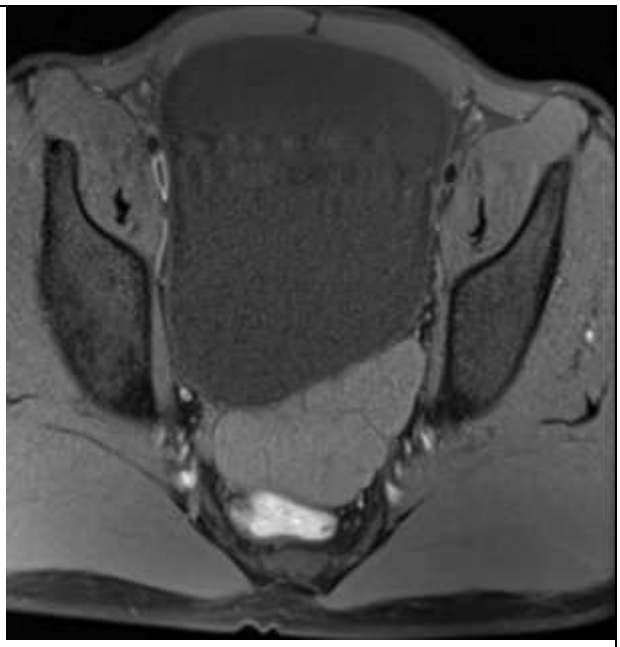

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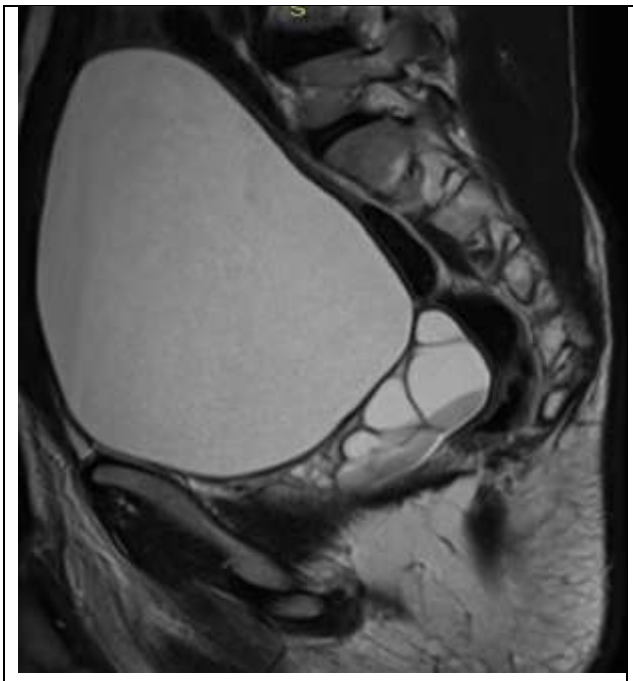

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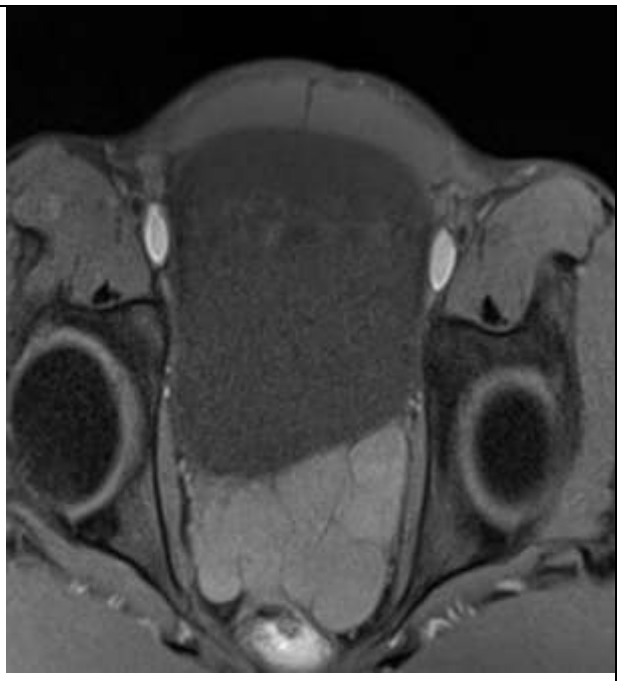

2.d

Fig 2. (a) MRI T2-W image revealing left renal agenesis; and s coronal (a) and T2-W and T1-W images in sagittal and axial planes revealing large ipsilateral left seminal vesicle cyst.

Case 3:

A 28-year-old male presented to the Urology OPD complaining of frequent urination that had been experienced over a month and accompanied by lower abdominal pain. Physical examination and CBC results were normal. Semen analysis showed azoospermia. The USG and CT showed absence of right kidney with an ipsilateral hypo-dense retro-vesical cystic structure. MRI was in agreement with the CT findings and revealed a unilateral cyst in the seminal vesicle with right renal agenesis (Fig.3). Diagnosis of Zinner syndrome was made and seminal vesicle cyst excision was accordingly scheduled.

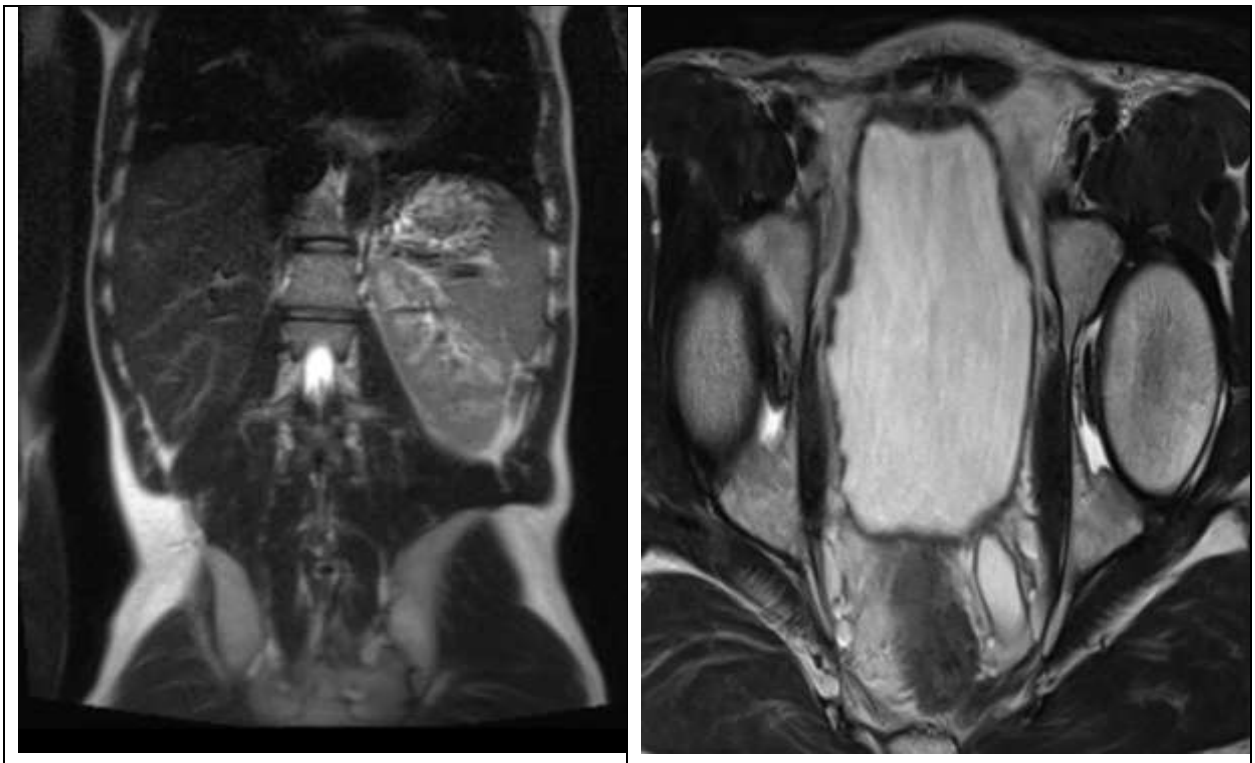

Image 3.a | Image 3.b

Fig 3. (a) MRI showing agenesis of the right kidney, with (b) contralateral left seminal vesicle cyst

Discussion

Ziner syndrome is a combination of renal agenesis, ejaculatory duct obstruction and ipsilateral cystic seminal vesicle; the condition was first described by Ziner in 1914 [1]. As per a large USG screening revealed that 13 out of 280,000 newborns in Taiwan exhibited cystic pelvic dilation with ipsilateral renal agenesis or dysplasia [6]. Another study reported that Zinner syndrome was observed in 214 cases per 100,000 patients [7]. Megaureter or ectopic ureter [8] are two associated anomalies frequently associated with this disorder.

The condition arises from a developmental abnormality of the mesonephric or Wolffian duct during weeks four to thirteen of pregnancy [9,10]. It has been described as the male equivalent of the female Mayer-Rokitansky-Kaiser-Hauser syndrome [11,12,13].

The urinary system primarily starts developing from the mesonephric duct. Growth factors released by the ureteric bud stimulate its growth toward the metanephric blastema [14,15]. The metanephric blastema fuses with the ureteric bud to produce the renal structure [16]. When this fusion process is disrupted, it leads to renal agenesis or renal hypoplasia. Between weeks six and eight of pregnancy, the ureteric bud separates from the distal end of the mesonephric duct [17]. This distal segment gives rise to seminal vesicle, vas deferens, ejaculatory ducts, hemitrigone,

bladder neck and urethra up to external sphincter, epididymis, paradidymis and appendix of epididymis [18].

In week twelve, the seminal vesicles are formed by the development of the penis mesonephric duct [3]. There is also the case whereby the ureteric bud cannot detach itself in the lower part of the mesonephric duct; this results in ejaculatory duct atresia and consequent seminal vesicle obstruction, leading to accumulation of the secretions thereby causing cystic dilation [7,11]. Furthermore, there also can be an ectopic ureter when the development of ureteric bud is not close enough to the urogenital sinus because too much time passes and the former cannot separate with the latter [19].

The condition is typically presents between the second and fourth decades of life. The patient frequently becomes aware of this issue while they are of reproductive age, either as a result of infertility or the onset of symptoms. It has been demonstrated, specifically, that the patient is likely to remain asymptomatic until the seminal vesicle cysts becomes about 5 cm in size. Urinary tract infections or inflammations, as well as mass effects on other organs can develop from large cysts.

The most prevalent symptoms of Zinner syndrome as reported by Van den Ouden and coworkers [20] in their paper analyzing symptomatology in 52 patients of Zinner syndrome include dysuria in 37%, urinary frequency in 33%, perineal discomfort in 29%, epididymitis in 27%, pain during ejaculation in 21% and scrotal pain in 13 percent. Symptoms frequently flare up and go away over time, occasionally with the use of non-steroidal anti-inflammatory drugs [21]. Two additional notable results of this study were the occurrence of ureteral remnants causing cysts in only 27% of patients and the higher frequency of abnormalities on the right side (ratio: R:L = 2:1). In contrast, two of our patients had ureteral remnant, and the affected side was the left.

In people of reproductive age, oligozoospermia and azoospermia can result in infertility [12,13]. Before beginning any diagnostic imaging investigations, it is imperative to do a proper physical examination on the patient. Rectal examination alone has been demonstrated to be enough for feeling the size of the cysts [20].

Owing to transabdominal USG being a noninvasive, low-cost and radiation-free imaging tool, it is the first diagnostic method to be employed [21,22], especially in younger patients. Transrectal USG can be employed especially in identifying seminal vesicle morphology and can be used in monitoring clinically asymptomatic seminal vesicle cyst. The typical features on sonography are those of an anechoic mass enclosing the seminal vesicles. It is essential to distinguish seminal vesicle cysts from other cystic conditions of the pelvic organs. The location of the cyst is a key factor in the differential diagnosis. Seminal vesicle cysts are typically positioned in the paramedian area, contrasting with prostatic duct and prostatic utricle cysts which are typically found along the midline. Unlike seminal vesicle cysts, which are usually round or oval with smooth borders and occur without associated renal agenesis, Zinner syndrome-related seminal vesicle cysts tend to be larger, more spherical or tubulo-saccular in shape and exhibiting irregular edges [2-4]. As with our first patient, there are times when the mass may be iso-hyperechoic,

which might turn out to be due to purulent material from an infection or the proteinaceous content of the vesicle [22].

Since USG is an operator-dependent modality, the next imaging tool to determine the cyst's connection to the structures surrounding is CT. A common CT finding is that of a retrovesical periprostatic cystic mass associated with unilateral renal agenesis. Kenney and Leeson found a variety of imaging outcomes in their study; a thick and irregular wall cystic pelvic mass, a well defined low density retrovesical mass whose origins were in the seminal vesicle or a mass adjacent to the prostate gland [22].

With advancements in multiplanar imaging technology and the availability of newer MR sequences, MRI has become the most effective technique for assessing pelvic masses and identifying any anatomical connections and morphological details with adjacent organs. Resultantly, MRI has become a valuable tool both for confirming the diagnosis and planning surgery, when removal of seminal vesicle cysts is contemplated [18].

The typical MRI findings of a seminal vesicle cyst include paramedian and periprostatic localization; diverse patterns of signals on T1-weighted scans and hyperintense signals on T2-weighted scans [23,24]. Although simple cysts in most cases would be hypointense on the T1-weighted images and hyperintense on T2-weighted images, depending on the amount of protein-rich or infected fluid, the intensity may be increased in the T1-weighted images. Giant cysts over 12 centimeters have also been reported in the literature restricting the bladder and bowels; however vast majority are small being less than 5 cm in size [25].

Conclusion

While Zinner syndrome is a rare congenital urogenital affection, yet the condition needs to be kept in mind when confronted with patients having typical symptomatology and clinical features. While sonography would continue to be the first line of investigation in such cases, yet cross-sectional multiplanar imaging tools provide an in-depth morphological insight into the problem. Availability of newer MRI sequences make it possible to reveal detailed anatomical mapping of the condition, which becomes invaluable for planning precise and minimally invasive surgical intervention.

Author Contributions

Ayesha Naaz: Writing - Original Draft, Writing – Review and editing.

Dr Mudasir: Writing, Writing – Review
and editing.

Dr Irfan Robbani: Responsible for drafting the original manuscript and contributing to its review and editing

Ethical Considerations

This case series was exempt from ethical committee approval as it involved retrospective anonymized data collection.

Funding:

The authors did not receive any funding for the preparation of this manuscript

Conflict of Interests:

There are no competing interests to declare.

Data availability statement:

Data supporting the findings of this study are available from the corresponding author upon reasonable request via email.

Ethical Statement:

This study was conducted in accordance with the ethical principles outlined in the Declaration of Helsinki (2013), as revised by the World Medical Association.

Patient consent:

Written informed consent was obtained from all patients for publication of their case details and associated images.

Acknowledgements:

This case report is based on cases managed at SKIMS Hospital, with clinical findings and outcomes verified by the Department of Surgery, SKIMS, Srinagar.

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