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FIVE DECADES OF SILENCE: DELAYED SYMPTOMATIC PRESENTATION OF ZINNER SYNDROME WITH ECTOPIC RENAL REMNANT AND MRI-OCCULT SEMINAL VESICLE CALCULI

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ABSTRACT **Background:** Zinner syndrome (ZS) is a rare congenital anomaly of the mesonephric (Wolffian) duct, defined by the triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst (SVC), and ipsilateral ejaculatory duct obstruction (EDO). The mean age of diagnosis across 214 pooled cases is 29.35 years; presentation beyond the fifth decade is exceptional. **Case Presentation:** A 55-year-old male presented with an 8-month history of lower urinary tract symptoms (LUTS) and right flank pain. MRI pelvis confirmed the complete Zinner triad: absent right kidney (ipsilateral renal agenesis), a 7×3×4 cm T2-hyperintense right seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction. A non-functional pelvic ectopic reniform structure (70×45 mm) was also identified on the right side. Seminal vesiculoscopy demonstrated multiple intraluminal calculi not identified on pre-operative imaging. **Management:** The patient underwent seminal vesiculoscopy followed by robot-assisted laparoscopic right seminal vesiculectomy with en-bloc remnant ureter excision. The postoperative course was uneventful. **Conclusion:** This case illustrates three compounding novelties: delayed symptomatic presentation of ZS at 55; a pelvic ectopic reniform remnant rather than true renal aplasia satisfying the ipsilateral renal agenesis arm of the triad; and intraoperative seminal vesicle calculi invisible on MRI. ZS should be suspected in any male with LUTS or perineal pain in whom imaging reveals an absent or anomalously positioned ipsilateral kidney, irrespective of age.

KEYWORDS : Zinner syndrome; seminal vesicle cyst; ipsilateral renal agenesis; ejaculatory duct obstruction

INTRODUCTION

A 55-year-old farmer from Muzaffarnagar arrived in our outpatient clinic with eight months of burning micturition and a dull ache in his right flank. Nothing in his presentation immediately suggested a congenital anomaly. Yet the imaging that followed would reveal a developmental error encoded in the fourth week of his embryonic life — one that had persisted silently for five and a half decades before finally making itself known.

Zinner syndrome is a rare anomaly of the mesonephric (Wolffian) duct arising from a focal developmental failure between gestational weeks 4 and 13. The disruption simultaneously produces three ipsilateral consequences: the kidney fails to form (**renal agenesis**), seminal fluid has nowhere to drain because the ejaculatory duct is atretic (**ejaculatory duct obstruction**), and back-pressure progressively distends the seminal vesicle into a cyst (**seminal vesicle cyst**). This triad — all three elements on the same side — is the diagnostic hallmark of Zinner syndrome.

Liu *et al.* analysed 214 published cases and established a mean age of diagnosis of 29.35 years.[1] The syndrome is typically unmasked by haematospermia, perineal pain, LUTS, or infertility in the second or third decade. At 55, our patient stands more than two standard deviations beyond that mean.

Three additional features distinguish this case: the ipsilateral renal anomaly proved to be a non-functional pelvic ectopic reniform remnant rather than true aplasia; multiple intraluminal seminal vesicle calculi were identified only on intraoperative vesiculoscopy; and the long-neglected cyst required perioperative complexity management in an older patient. Together these constitute a constellation not previously described in the Indian urological literature.

CASE REPORT

Presentation. A 55-year-old male was admitted on 18 May 2026 with lower urinary tract symptoms including burning micturition of 8 months 'duration and right flank pain for the preceding month. He denied haematospermia, perineal pain, or any prior urological workup. He had a background of hypertension and coronary artery disease (PTCA 2015) and was on amlodipine, metoprolol succinate, and aspirin-atorvastatin.

Examination. He was conscious and haemodynamically stable (HR 63/min, BP 137/90 mmHg, SpO_2 99% on room air, afebrile). Abdominal examination was unremarkable except for mild suprapubic tenderness. Digital rectal examination revealed a Grade 1 prostate, smooth and non-nodular.

Investigations. Haemoglobin was 11.7 g/dL at admission (rising to 13.2 g/dL pre-operatively). Renal function was preserved with a stable creatinine of 1.11–1.13 mg/dL, consistent with compensatory hypertrophy of the solitary contralateral left kidney. Mild hyponatraemia (Na^+ 130 mEq/L) was noted and monitored. Urine culture was sterile on both occasions (Table 1).

Table 1: Laboratory investigations

Investigation	Value
Haemoglobin (g/dL)	11.7 (admission); 13.2 (pre-operative)
TLC ($\times 10^3/\mu L$)	7.54
Platelets ($\times 10^3/\mu L$)	193
Urea / Creatinine (mg/dL)	18 / 1.11
Na^+ / K^+ (mEq/L)	130 / 4.2
T. Bil / D. Bil (mg/dL)	0.88 / 0.32
SGPT / SGOT / ALP (U/L)	26 / 34 / 122
PT / INR	1.1
HIV / HCV / HBsAg	Non-reactive
Urine C/S	Sterile

Imaging — CECT abdomen and pelvis. A contrast-enhanced CT performed prior to referral showed the right kidney to be absent from

its normal retroperitoneal position (ipsilateral renal agenesis). A reniform structure measuring 70×45 mm was identified in the right pelvis adjacent to the bladder wall, without enhancement or contrast excretion in the delayed phase — a non-functional pelvic ectopic remnant on the same side as the absent kidney. The left kidney measured 97×58 mm with mild perinephric fat stranding and multiple small non-enhancing cortical cysts. Importantly, the right seminal vesicle cyst was not characterised on CT, underscoring CT's well-recognised limitation in soft-tissue pelvic detail.

Imaging — MRI pelvis with abdomen. MRI confirmed all three elements of the Zinner triad on the right side (Figures A–D). First, the right kidney was completely absent from the retroperitoneum; coronal sequences showed the right renal fossa occupied only by liver, with no renal parenchyma below it (Figures A, C). The right adrenal gland was flattened and elongated on axial sequences (the 'lying-down adrenal sign', confirmatory of ipsilateral renal agenesis). Second, a well-defined cystic lesion measuring 7×3×4 cm occupied the right retrovesical space, strikingly T2-hyperintense (Figures A, B, C) and T1-hypointense (Figure D), with no internal enhancement or diffusion restriction — the right seminal vesicle cyst. Third, the ejaculatory duct on the right side was not patent, consistent with ipsilateral ejaculatory duct obstruction completing the triad. The MRI conclusion read: 'right renal agenesis with ipsilateral seminal vesicle cysts, consistent with Zinner syndrome.'

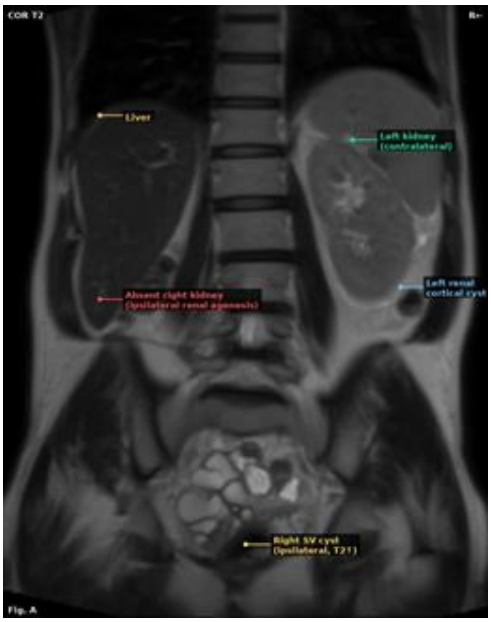

Figure A: Coronal T2 MRI: Liver occupies the right upper quadrant (viewer's left, R←). The right renal fossa is entirely empty (ipsilateral renal agenesis) — Arm 1 of the Zinner triad. The contralateral left kidney with a cortical cyst is seen in the left retroperitoneum. The right SV cyst (ipsilateral, T2-hyperintense) is visible in the inferior pelvis — Arm 2 of the triad. Ejaculatory duct obstruction (Arm 3) is confirmed on MRI as a non-patent right ejaculatory duct.

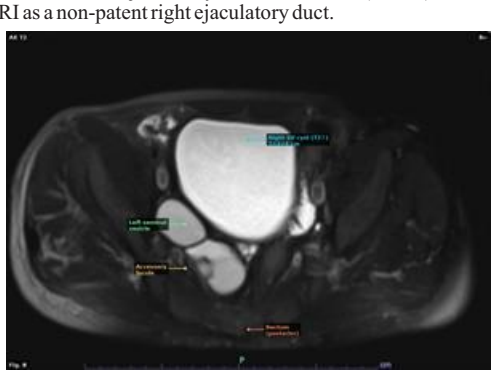

Figure B: Axial T2 MRI at the level of the seminal vesicles: The right SV cyst (7×3×4 cm, ipsilateral) is strikingly T2-hyperintense. The contralateral left seminal vesicle is identified posteriorly as a smaller grey structure. An accessory locule is visible at the inferior margin of the cyst. The rectum lies posterior.

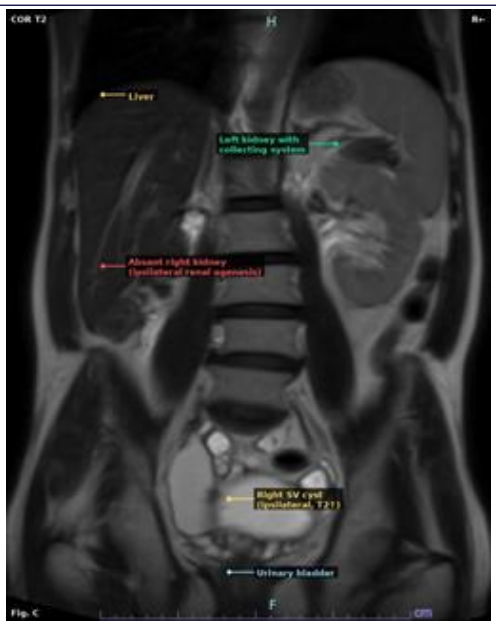

Figure C: Coronal T2 MRI at a more posterior plane: The left kidney with its intrarenal collecting system is clearly demonstrated (contralateral). On the patient's right side (viewer's left), the renal fossa is completely empty — no renal parenchyma is present (ipsilateral renal agenesis). The ipsilateral right SV cyst and the urinary bladder are well demonstrated in the inferior pelvis, illustrating all three elements of the Zinner triad.

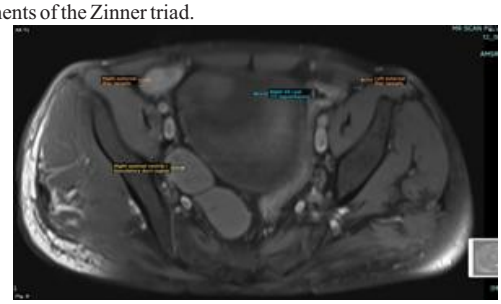

Figure D: Axial T1 MRI pelvis: The right SV cyst is T1-hypointense (simple fluid, no haemorrhage). The right seminal vesicle and ejaculatory duct region are labelled, where obstruction of the ejaculatory duct — the third arm of the triad — was confirmed. Bilateral external iliac vessels serve as anatomical landmarks.

Surgical management — Stage 1: Seminal vesiculoscopy. Formal anaesthetic clearance was obtained and antiplatelet therapy managed perioperatively. With the patient in low lithotomy position, a 7.5/8 Fr ureteroscope was advanced transurethrally. The urethra, prostatic fossa, and bladder neck were normal. The prostatic utricle was entered and the right seminal vesicle orifice cannulated. Within the right SV lumen, multiple calculi were encountered — entirely unexpected on MRI, and a clinically significant additional finding. The left ureteric orifice was identified at the normal trigonal position; the right ureteric orifice was absent, consistent with the ipsilateral renal agenesis.

Surgical management — Stage 2: Robot-assisted laparoscopic vesiculectomy. Pneumoperitoneum was established and ports placed. The posterior peritoneum was incised and posterior dissection performed. Bilateral vasa deferentia were identified as landmarks. The right SV cyst (approximately 7×3 cm) was dissected from surrounding structures; the ipsilateral ectopic ureter was traced to the iliac vessel crossing and divided between Hem-o-lok clips. The SV was separated distally at the ejaculatory duct level and the right vas deferens cauterised and ligated. The specimen was retrieved via the 11 mm port, a 20 Fr drain placed, and the abdomen closed in layers.

Postoperative course. The drain and catheter were removed on POD 1. The patient mobilised on POD 2 and was discharged on POD 5 in stable condition. He was prescribed co-trimoxazole DS for one month and reviewed at one week for suture removal. Histopathology (G-1849/26, AIIMS Rishikesh Department of Pathology and Lab

Medicine, reported 26 May 2026) confirmed the diagnosis. Gross examination of the excised cyst (4.1×2.8×1.1 cm) revealed a multiloculated cavity containing golden-brown material; the separately received seminal vesicle measured 5×4×2 cm and the attached ureteric segment was 6.2 cm in length. On microscopy, the cyst lining showed cuboidal to columnar epithelium without atypia; glandular spaces contained intraluminal eosinophilic granular secretions. The seminal vesicle showed no significant pathology. No evidence of atypia or granulomatous inflammation was identified. These findings are consistent with a benign seminal vesicle cyst of Wolffian duct origin, with no malignant transformation.

DISCUSSION

The three arms of the Zinner triad arise from a single embryological error in the mesonephric duct between weeks 4 and 13 of gestation. When the ureteric bud fails to migrate correctly from the proximal portion of the mesonephric duct, it cannot induce metanephric differentiation, resulting in ipsilateral renal agenesis. The same duct abnormality distally produces atresia of the ejaculatory duct, which in turn prevents drainage of seminal fluid and causes progressive distension of the seminal vesicle into a cyst. All three consequences are ipsilateral — this is the defining anatomical logic of the syndrome.[1,2]

The most striking epidemiological feature is timing. ZS is overwhelmingly a disease of young men;fewer than 3% of 214 pooled cases were diagnosed beyond age 45.[1] Our patient was 55. The explanation lies in the mechanics of obstruction: ejaculatory duct atresia is congenital, but its consequence — seminal vesicle distension — is cumulative. Seminal fluid production begins at puberty and the vesicle slowly enlarges over decades, compressing the bladder only when it reaches a critical volume. In this patient, that threshold was reached at 55.

The ipsilateral renal anomaly in this case merits careful classification. CECT identified a 70×45 mm non-functional reniform pelvic structure on the right side — the same side as the absent retroperitoneal kidney and the seminal vesicle cyst. The absence of corticomedullary enhancement or contrast excretion, together with the ipsilateral 'lying-down adrenal sign 'on MRI, supports renal aplasia with persistence of a vestigial mesonephric pelvic remnant, rather than a normally structured ectopic kidney.[3] Functionally and for the purposes of the Zinner triad, the right renal fossa was empty and the contralateral left kidney was the sole functioning unit.

The intraoperative discovery of seminal vesicle calculi is a noteworthy finding invisible on MRI. Stasis calculi within an obstructed seminal vesicle form by mineral precipitation in a chronically static, protein-rich fluid column — directly analogous to stones in an obstructed ureter. Their reported incidence in ZS is under 5%.[4] This case reinforces that vesiculoscopy is not merely therapeutic but provides diagnostic information that even gold-standard MRI cannot.

The treatment choice — robotic vesiculectomy with en-bloc remnant ureter excision — was driven by the clinical profile: a 55-year-old with no fertility aspiration, in whom the ejaculatory duct obstruction had already caused irreversible seminal vesicle enlargement. Transurethral resection of the ejaculatory duct (TURED), appropriate in younger men seeking fertility restoration, was not indicated.[1] Complete excision eliminated the risks of future cyst infection (documented as potentially lethal in the ZS literature) and malignant transformation, reported in approximately 4.4% of cases.[5]

CONCLUSION

Zinner syndrome is a congenital anomaly of the mesonephric duct that may remain clinically silent for decades, manifesting only when progressive seminal vesicle distension reaches a critical volume. This case adds three instructive novelties to the literature: delayed symptomatic presentation of a congenital anomaly at 55; a pelvic ectopic reniform remnant satisfying the renal agenesis arm of the triad; and seminal vesicle calculi discoverable only on intraoperative vesiculoscopy. ZS should be suspected in any male presenting with LUTS or perineal pain in whom imaging incidentally reveals an absent or anomalously positioned ipsilateral kidney, irrespective of age. Pelvic MRI confirms the triad; vesiculoscopy detects what imaging misses; robot-assisted laparoscopic vesiculectomy provides definitive, safe treatment.

DECLARATIONS

Patient consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Ethics: This report complies with institutional ethical standards. Patient identity has been protected throughout.

Conflicts of interest: None declared.

Funding: None.

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