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Zinner Syndrome: A Case Report

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Abstract: Zinner syndrome is a rare congenital anomaly resulting from mesonephric duct maldevelopment and characterized by the triad of ipsilateral renal agenesis, seminal vesicle cyst(s), and ejaculatory duct obstruction. We report the unique case of an 18-year-old man with a history of solitary right kidney and prior left varicocele ligation, who presented with left scrotal pain and swelling. This case represents a rare presentation of Zinner syndrome with both multiloculated and hemorrhagic seminal vesicle cysts. It underscores the importance of multimodal imaging and histopathological evaluation in diagnosing atypical variants of Zinner syndrome and guiding surgical intervention.

Keywords: *mesonephric duct anomaly, renal agenesis, robotic vesiculectomy, seminal vesicle cyst, Zinner syndrome*

Introduction

Zinner syndrome (ZS) is a rare congenital anomaly caused by maldevelopment of the mesonephric duct and classically presents with the triad of ipsilateral renal agenesis, seminal vesicle cysts, and ejaculatory duct obstruction.¹ Although ZS is often asymptomatic until early adulthood, patients may present with pelvic pain, infertility, or genitourinary complaints.² Fewer than 300 cases have been reported, and hemorrhagic seminal vesicle cysts represent an exceedingly rare manifestation.^{3,4} Recent literature emphasizes the role of multimodal imaging, particularly MRI, in diagnosis.⁵ This case describes an 18-year-old man with hemorrhagic, multiloculated seminal vesicle cysts associated with Zinner syndrome. We aim to expand the clinical spectrum of ZS and underscore the importance of recognizing atypical imaging features. This case report was prepared following the CARE Guidelines.⁶

Key Points

- Multimodal imaging is critical for diagnosis and surgical planning in cases of Zinner syndrome, but MRI most clearly images the entity's characteristic features of ejaculatory duct obstruction, seminal vesicle cysts, and renal agenesis.
- Laparoscopic surgery is the gold standard in the treatment of Zinner syndrome, resolving pain and potentially improving fertility.
- Clinicians should suspect Zinner syndrome in patients with renal agenesis even when cystic pelvic findings are atypical.

Case Presentation

An 18-year-old man presented to a urologist with complaints of left-sided scrotal pain and swelling. One week prior, the patient had undergone surgical ligation of a varicocele on the left side of his scrotum. Two months prior to the varicocele

Abbreviations

AFP: α -fetoprotein

β -hCG: β -human chorionic gonadotropin

CF: cystic fibrosis

FSH: follicle-stimulating hormone

LDH: lactate dehydrogenase.

LH: luteinizing hormone

TURED: transurethral resection of the ejaculatory duct

US: ultrasonography

ZS: Zinner Syndrome

ligation, scrotal ultrasound revealed tubular ectasia of the rete testis as well as significant chronic enlargement of the left testicle that may or may not have been due to a deflated varicocele. The patient's medical history was also significant for a solitary right kidney.

Physical examination for the patient's left-sided scrotal pain and swelling revealed that the left hemiscrotum was significantly enlarged compared to the right. The vas deferens on the left side was not palpable. Scrotal ultrasound (Figure 1) showed a moderately enlarged left testicle containing a tubular hypoechoic structure with little to no Doppler flow, representing severe tubular ectasia of the rete testis with complex internal contents. A mildly thick-walled, tortuous vas deferens was identified laterally. There was a trace left hydrocele, and no varicocele was seen. The right testicle and epididymis were unremarkable. Laboratory investigations, including genetic testing for cystic fibrosis and tests of endocrine hormone labs such as testosterone, estradiol, follicle-stimulating hormone (FSH), luteinizing hormone (LH), α -fetoprotein (AFP), β -human chorionic gonadotropin (β -hCG), and lactate dehydrogenase (LDH) returned within normal limits or negative. Semen analysis revealed slightly low sperm concentration, but all other parameters were normal.

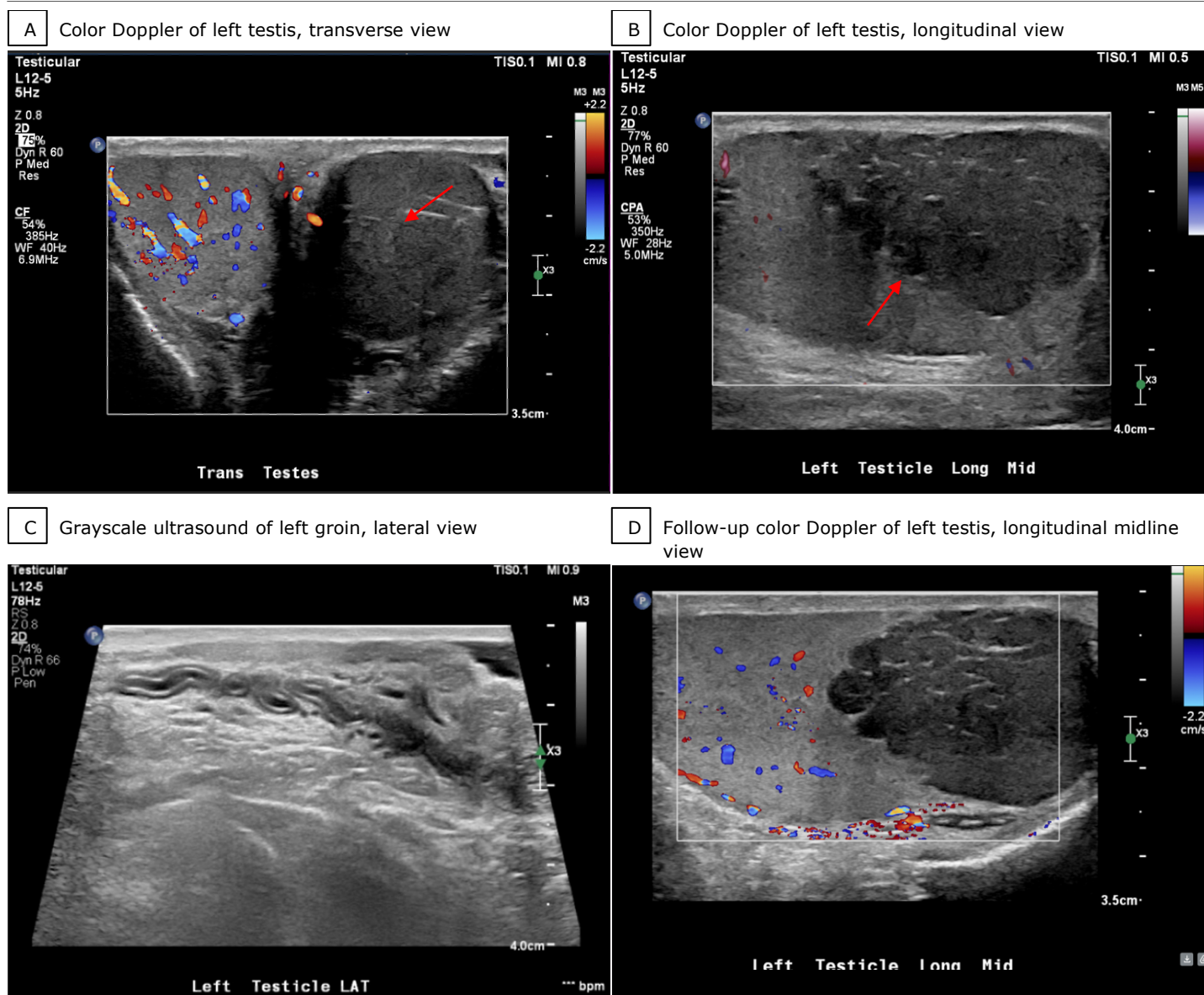
Three months after this examination, the patient developed abdominal pain, prompting further imaging. MRI of the pelvis (Figure 2) revealed a nonenhancing, multiloculated, T1-hyperintense dilation—likely from hemorrhagic or proteinaceous material—of the rete testis and the left seminal vesicle with absence or atresia of the left ejaculatory duct. Additionally, ipsilateral renal agenesis was confirmed. Follow-up ultrasound one

month later better showed blood flow to parts of the left testis on Doppler ultrasound (Figure 1D). The following month, the patient underwent cystoscopy, which revealed an enlarged verumontanum and deformation of the left lateral trigone. During the procedure, 200 cc of fluid was drained from the left seminal vesicle, providing symptomatic relief. For definitive management, the patient was offered transurethral resection of the ejaculatory duct (TURED), repeat percutaneous aspiration, and seminal vesiculectomy. Ultimately, he underwent a robotic left seminal vesiculectomy. Intraoperatively, the left seminal vesicle was traced to the level of the prostate, where no visible lumen was identified. The left seminal vesicle and the left vas deferens were completely occluded. Histopathologic examination of the left seminal vesicle revealed a 42 g partially disrupted and cauterized fragment of seminal vesicle with hemorrhagic fluid and no lesions or other abnormalities.

Discussion

Zinner syndrome results in a rare cystic malformation of the seminal vesicle that occurs due to abnormalities in distal mesonephric duct development during embryogenesis. This process takes place between the 4th and 13th weeks of gestation. The inability of the ureteric bud to fully migrate and initiate the development of the metanephros is the pathophysiological basis of ZS and leads to atresia of the ejaculatory duct and ipsilateral renal agenesis. Because of the ejaculatory duct atresia, fluid cannot adequately drain, leading to the characteristic cystic dilatation of the ipsilateral seminal vesicle.⁶ The abnormal development results in the classic triad of unilateral renal agenesis (or in some cases multicystic dysplastic kidney), cystic dilatation of the ipsilateral seminal vesicle, and ipsilateral ejaculatory duct obstruction.⁵ Due to their similar pathophysiologies, ZS is commonly recognized as the male counterpart of Mayer-Rokitansky-Küster-Hauser syndrome, also known as Müllerian agenesis.⁸

Zinner syndrome is often asymptomatic until the second or third decade of life, when patients

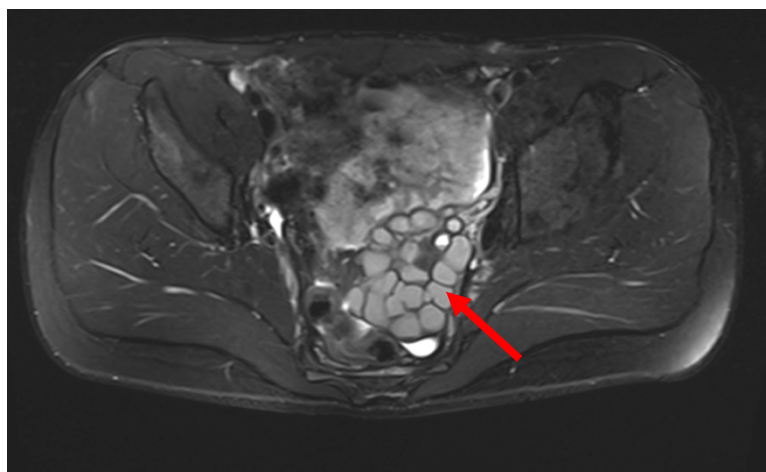
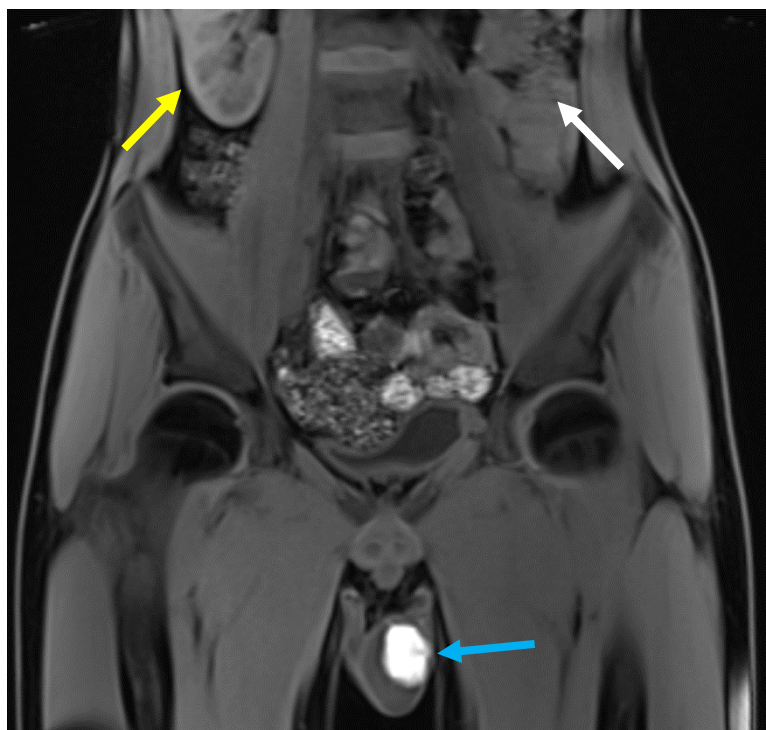
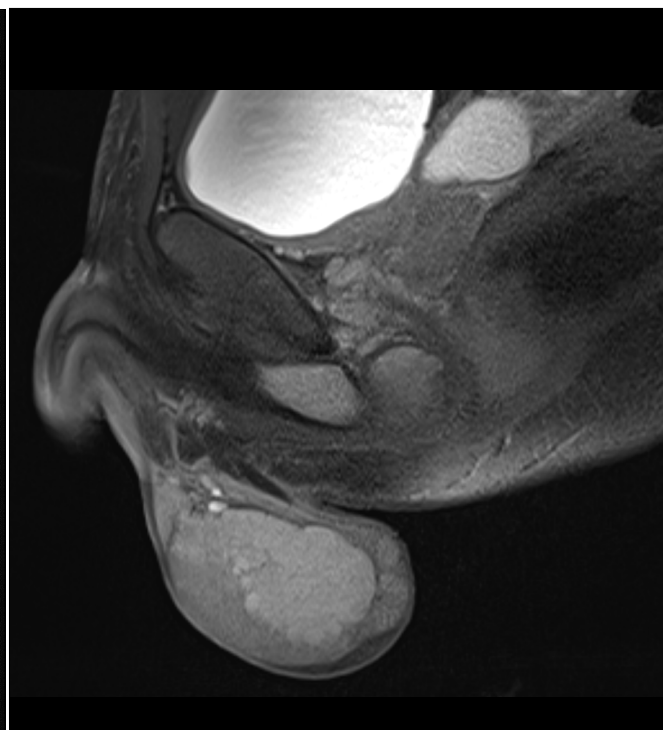
Figure 1. Scrotal Ultrasound in an 18-Year-Old Man with Zinner Syndrome

(A) Transverse and (B) longitudinal Color Doppler ultrasound show a moderately enlarged left testis (5.5 × 3.2 × 4.1 cm) (red arrow), containing a lobular hypoechoic area (4.5 × 2.6 × 2.9 cm) with little to no Doppler flow, indicating dilation of the rete testis. (C) Grayscale ultrasound of the left groin shows a dilated vas deferens. (D) Follow-up color Doppler of left testis in longitudinal midline view better demonstrates flow to the left testis four months after initial ultrasound.

typically become sexually active.⁹ In rare cases it has been diagnosed as early as infancy, with around 13 cases reported during infancy and around 50 cases in the pediatric population as of 2022.⁹ When cases become symptomatic, complaints tend to include vague scrotal, perineal, abdominal, or pelvic pain.² This pain is usually exacerbated by defecation and ejaculation. Rarely, patients can also present with hematospermia.¹⁰ Infertility can be present in up to 45% of patients

with ZS, primarily due to congenital ejaculatory duct obstruction.² This case is unique in that the patient presented not only with hemiscrotal pain but also with hemiscrotal swelling, which prompted ultrasound imaging.

When Zinner syndrome is suspected, an MRI of the pelvis is the gold standard for diagnosis, as it shows the characteristic seminal vesicle cyst(s) and ejaculatory duct atresia or obstruction.⁵ In this case, the initial study ordered was a scrotal

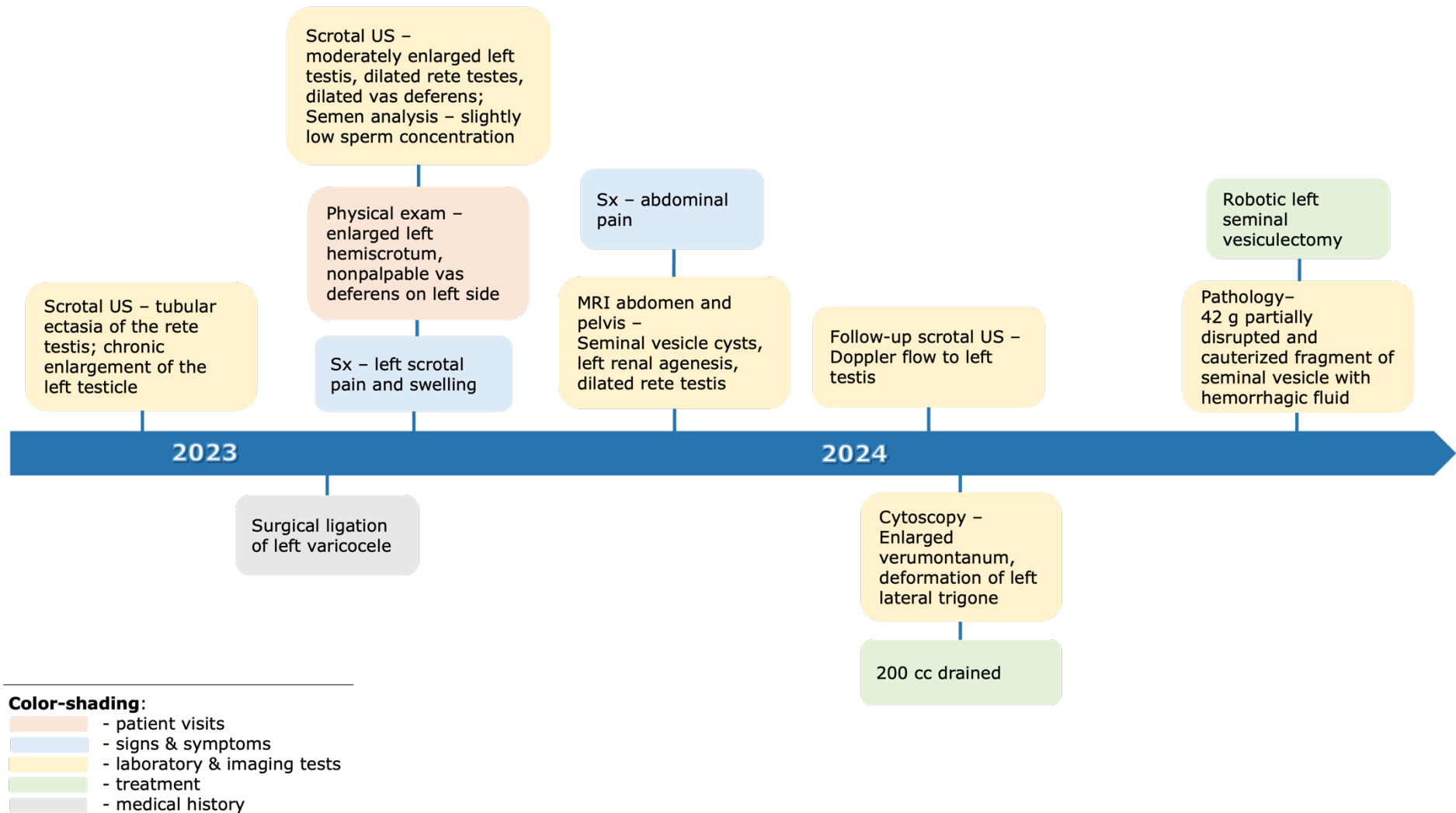
Figure 2. Magnetic Resonance Images of an 18-Year-Old Man with Zinner Syndrome**A** T2-weighted, fat-saturated MRI of the pelvis, axial view**B** T1-weighted, fat-saturated MRI of the pelvis, coronal view**C** T1-weighted, fat-saturated MRI of the abdomen and pelvis, coronal view**D** Postcontrast T1-weighted, fat-saturated MRI of the left testicle, sagittal view

(A) Axial T2-weighted fat-saturated and (B) coronal T1-weighted fat-saturated images demonstrate T1- and T2-hyperintense dilation of the left seminal vesicle and seminal vesicle cysts (red arrow). (C) A coronal T1-weighted fat-saturated image shows a T1-hyperintense dilated rete testis in the left testicle (blue arrow), absence of the left kidney in the lower abdomen (white arrow), and a partially visualized right kidney demarcated (yellow arrow). (D) A sagittal postcontrast T1-weighted fat-saturated image of the left testicle shows a lack of significant enhancement within the left testicular lesion.

ultrasound (Figure 1), as the patient presented with hemiscrotal swelling and a nonpalpable vas deferens one week after surgical varicocele ligation. MRI of the abdomen and pelvis was

prompted later by the occurrence of abdominal pain (Figure 2). Abdominal MRI of a patient with ZS typically shows ipsilateral renal agenesis or, less commonly, a multicystic dysplastic kidney.¹¹

Case report timeline



Abbreviations: MRI, magnetic resonance imaging; US, ultrasound; Sx, symptoms

Seminal vesicle cysts typically appear with low signal intensity on T1-weighted images and high attenuation on T2-weighted images.³ In the case of our patient, the multiloculated cyst in the left testis showed T1 hyperintensity, indicating the presence of hemorrhage or proteinaceous content within the cyst. Pathologic examination later confirmed the presence of hemorrhagic fluid.

Based on our literature search, hemorrhagic cysts in the setting of Zinner syndrome are a rare finding in an already rare disease. Cases showing T1-hyperintense, T2-hypointense lesions are very rare, with the most recent case being Xing et al in 2022, which was the first reported case of a single giant seminal vesicle cyst with hemorrhage in ZS.⁴ This case differs due to the presence of multiple hemorrhagic cysts.

Asymptomatic patients with ZS are managed conservatively with regular follow-up to monitor for progression, even if they are diagnosed in infancy.¹¹ Mildly symptomatic patients or patients who prefer noninvasive interventions can also be treated with α 1-adrenoceptor antagonists to alleviate ejaculatory and pelvic pain.¹⁰ Definitive management involves surgical excision of the seminal vesicle cyst(s).⁷

Conclusion

This case highlights a rare presentation of Zinner syndrome with multiloculated and hemorrhagic or proteinaceous seminal vesicle cysts and scrotal pain and swelling. The hemorrhagic or proteinaceous nature likely reflects chronic obstruction with superimposed bleeding or concentration of seminal vesicle contents. For urologists and radiologists, this case emphasizes the value of multimodal imaging in diagnosis and surgical planning, while pathology provides definitive confirmation. In this case, robotic vesiculectomy offered definitive relief and may preserve long-term reproductive potential.² Due to the long-term infertility risk associated with ZS, there is great clinical relevance in timely diagnosis and intervention. From a teaching standpoint, this report expands the clinical spectrum of ZS, reminding clinicians to suspect it in adolescents with renal agenesis and unusual cystic pelvic findings.

Author Contributions

Conceptualization, M.S.T., M.M., and M.K.P.; Acquisition, analysis, and interpretation of data, M.S.T.; Writing—original draft preparation, M.S.T.; Review and revisions, M.S.T., M.M., and M.K.P.; Supervision, M.M. and M.K.P. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Disclosures

None to report.

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