

Radiological Case Report / Caso Clínico

Zinner Syndrome in Adolescence: Imaging Findings

*Síndrome de Zinner na Adolescência: Achado Imagiológicos*Fabiana Muñoz¹, Teresa Tavares², Teresa Costa², Filipa Lima Coelho²¹Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal²Unidade Local de Saúde de Santo António, Porto, Portugal

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Abstract

Zinner syndrome is a rare congenital anomaly of the mesonephric duct characterised by the triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. The diagnosis is often made in early adulthood, although many cases remain asymptomatic and are detected incidentally. Imaging plays a crucial role in diagnosis, as clinical manifestations are often nonspecific. Ultrasound is typically the first-line modality, enabling detection of renal agenesis and retrovesical cystic lesions, while magnetic resonance imaging is considered the gold standard for its superior soft-tissue characterisation and multiplanar capability, allowing accurate depiction of seminal vesicle cysts and associated anomalies. We report the case of an asymptomatic 17-year-old male in whom imaging demonstrated right renal agenesis associated with an ipsilateral seminal vesicle cyst and a communicating ureteric remnant, consistent with Zinner syndrome. This case highlights the importance of imaging in recognising the characteristic features of this rare condition.

Keywords

Zinner syndrome; Renal agenesis; Seminal vesicle cyst; Ultrasonography; Magnetic resonance imaging.

Resumo

A síndrome de Zinner é uma anomalia congénita rara do ducto mesonéfrico caracterizada pela tríade de agenesia renal unilateral, quisto da vesícula seminal ipsilateral e obstrução do ducto ejaculatório. O diagnóstico é frequentemente estabelecido no início da idade adulta, embora muitos casos permaneçam assintomáticos e sejam detetados de forma incidental. A imagiologia desempenha um papel crucial no diagnóstico, uma vez que as manifestações clínicas são frequentemente inespecíficas. A ecografia é geralmente a modalidade de primeira linha, permitindo a deteção de agenesia renal e de lesões quísticas retrovesicais, enquanto a ressonância magnética é considerada o método de referência devido à melhor resolução de tecidos moles e capacidade multiplanar, permitindo a adequada identificação de quistos da vesícula seminal e de anomalias associadas. Apresenta-se o caso de um doente do sexo masculino, com 17 anos, assintomático, no qual os exames de imagem demonstraram agenesia renal direita associada a um quisto da vesícula seminal ipsilateral e remanescente ureteral comunicante, achados compatíveis com síndrome de Zinner. Este caso destaca a importância da imagiologia no reconhecimento das características típicas desta condição rara.

Palavras-chave

Síndrome de Zinner; Agenesia renal; Quisto da vesícula seminal; Ecografia; Ressonância magnética.

Introduction

Zinner syndrome is a rare congenital malformation of the mesonephric (Wolffian) duct characterised by the classical triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. It results from abnormal embryological development of the distal mesonephric duct during early pregnancy, leading to failure of ureteric bud formation with consequent renal agenesis and distal duct obstruction, causing cystic dilatation of the seminal vesicle.^{1,2}

Clinical presentation is variable and often non-specific, including lower urinary tract symptoms, pelvic or perineal pain, haemospermia, painful ejaculation, and infertility. However, many cases remain asymptomatic and are detected incidentally during imaging studies performed for unrelated reasons.^{1,3}

Imaging plays a central role in diagnosis. Ultrasound is usually the initial modality, allowing detection of renal agenesis and retrovesical cystic lesions, while magnetic resonance imaging (MRI) is considered the reference technique due to its superior soft-tissue contrast and multiplanar capability, enabling accurate characterisation of seminal vesicle cysts and associated pelvic anomalies.^{4,5}

Case Report

A 17-year-old male patient was referred to our institution following the incidental ultrasonographic diagnosis of unilateral renal agenesis, without associated symptoms. Ultrasound performed at our hospital confirmed right renal agenesis, with no evidence of an ectopic kidney, and demonstrated compensatory hypertrophy of the left kidney. In the pelvis, a well-defined cystic lesion was identified in the retrovesical periprostic region, apparently arising from

the right seminal vesicle. An adjacent tubular cystic structure extending along the right paravesical region was also noted, suggesting possible communication with the seminal vesicle cyst (Figure 1a). Further evaluation with MRI was therefore performed for detailed anatomical characterization and also confirmed the absence of the right kidney (Figure 2).

Pelvic MRI demonstrated enlargement of the right seminal vesicle due to cystic dilatation of its tubular components. A well-circumscribed cystic lesion was identified within the right seminal vesicle, located in the typical retrovesical paramedian position, posterior to the bladder and lateral to the prostate gland. A dilated tubular structure was also observed extending superiorly in the right paravesical region, compatible with a remnant ureter communicating with the seminal vesicle cyst (Figures 1b and 3).

The cystic structures demonstrated high signal intensity on T1-weighted sequences, both with and without fat suppression, and marked hyperintensity on T2-weighted sequences, findings consistent with proteinaceous or haemorrhagic content. The lesions were well defined, without internal septations or fluid–fluid levels. No contrast-enhanced sequences were obtained. The left kidney demonstrated normal morphology and signal intensity with preserved corticomedullary differentiation (Figure 2b).

Overall, the imaging findings were consistent with right renal agenesis associated with an ipsilateral seminal vesicle cyst and obstruction of the ipsilateral ejaculatory duct, in keeping with Zinner syndrome.

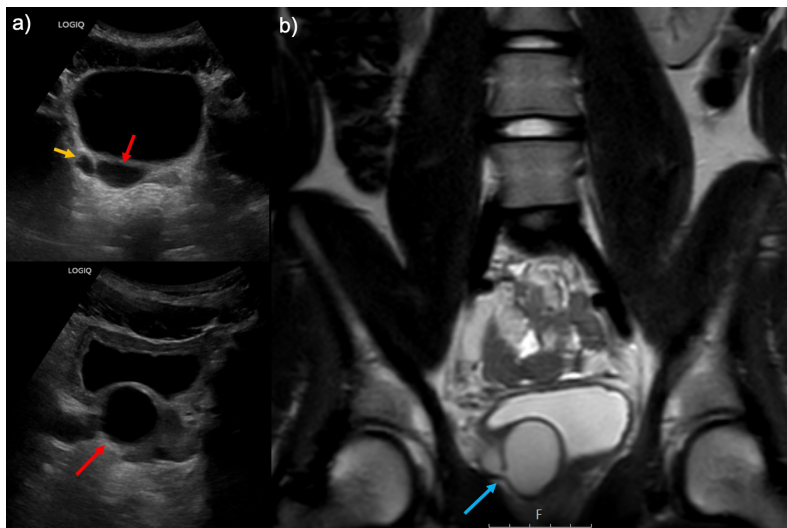


Figure 1 – a) Pelvic ultrasound: 3 cm cyst in the anatomical location of the right seminal vesicle (red arrow). Right paravesical dilated tubular structure leading to the described cyst (yellow arrow). b) Pelvic MRI: coronal T2-weighted sequence showing the connection between the cyst and remnant ureter.

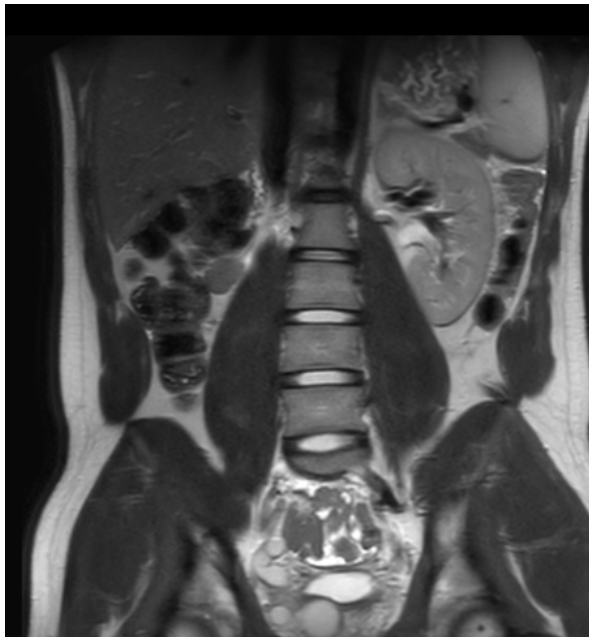


Figure 2 – T2-weighted MRI coronal section confirming the absence of the right kidney.

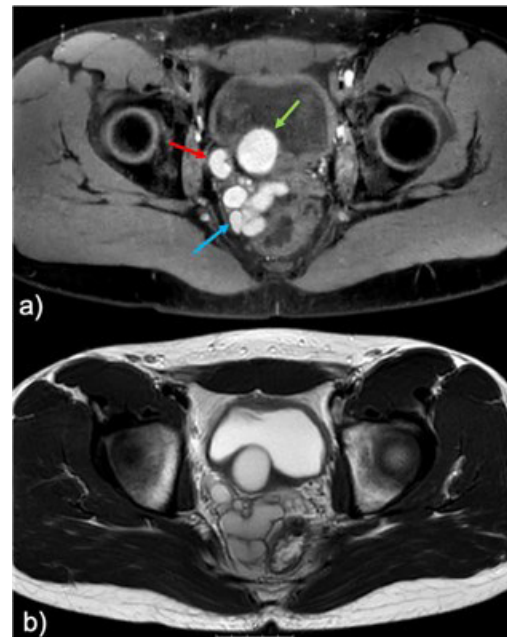


Figure 3 – Pelvic MRI a) axial fat-suppressed T1-weighted and b) axial T2-weighted sequences: enlarged right seminal vesicle, due to the dilatation of the tubular structures that form it (blue arrow). Right seminal vesicle cyst (green arrow) that communicates with the seminal vesicle ducts, as well as with a right paravesical dilated tubular structure compatible with rudimentary ureter (red arrow). These structures are hyperintense in T1-weighted and T2-weighted sequences, in relation to proteinaceous/haemorrhagic content.

Discussion

Zinner syndrome is a rare congenital malformation of the mesonephric (Wolffian) duct characterised by the classical triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and obstruction of the ejaculatory duct. First described by Zinner in 1914, it represents the male counterpart of Mayer–Rokitansky–Küster–Hauser syndrome in females, as both conditions arise from abnormalities in the development of mesonephric-derived structures. The syndrome results from abnormal embryological development of the distal mesonephric duct during early pregnancy, typically between the 4th and 13th weeks of embryonic life. Failure of ureteric bud formation results in renal agenesis, while distal obstruction of the ejaculatory duct leads to progressive accumulation of seminal secretions and cystic dilatation of the seminal vesicle. In some cases, a rudimentary ureter may persist and communicate with the seminal vesicle cyst, reflecting the shared embryological origin of these structures from the mesonephric duct.^{1,2}

Zinner syndrome is considered an uncommon condition, with only a limited number of cases described in the literature. A pooled analysis of 214 cases reported between 1999 and 2020 confirmed its rarity and highlighted the heterogeneity of clinical presentation. Most patients are diagnosed during the second or third decade of life, when increased seminal vesicle secretory activity following sexual maturation may lead to progressive cyst enlargement and symptom onset. Nevertheless, a substantial proportion of cases remain asymptomatic and are detected incidentally during imaging studies performed for unrelated reasons.^{1,3}

Clinical manifestations are often non-specific and may include dysuria, urinary frequency, pelvic or perineal pain, haemospermia, painful ejaculation and infertility. Symptoms are usually related to ejaculatory duct obstruction and mass effect from an enlarging seminal vesicle cyst on adjacent pelvic structures such as the bladder, prostate or rectum.^{1,5} Infertility may occur due to impaired seminal fluid drainage and altered sperm transport.¹ However, the correlation between cyst size and symptom severity is variable, and some patients remain asymptomatic even with relatively large cysts.¹

Imaging plays a pivotal role in diagnosis, as clinical findings are frequently subtle and the underlying anatomical abnormalities involve deep pelvic structures that cannot be adequately assessed clinically. The diagnostic workup should confirm the presence of renal agenesis, identify the seminal vesicle cyst, and assess associated anomalies involving the ejaculatory duct, vas deferens, or ureteric remnants, thereby providing essential information for clinical management and therapeutic planning.^{4,5}

Ultrasound is usually the first-line imaging modality and may demonstrate unilateral renal agenesis together with a retrovesical cystic lesion in the region of the seminal vesicle. Transabdominal ultrasound enables assessment of the kidneys and detection of pelvic cystic lesions, while transrectal ultrasound may provide a more detailed evaluation of seminal vesicle morphology and ejaculatory duct involvement.^{4,5} However, ultrasound may be limited in precisely defining the anatomical origin of retrovesical cysts and their relationship with adjacent pelvic structures. MRI is therefore considered the imaging modality of choice

due to its superior soft-tissue contrast and multiplanar capability. MRI allows accurate identification of seminal vesicle cysts, assessment of their anatomical relationships with the bladder, prostate, and vas deferens, and detection of associated abnormalities such as dilated ejaculatory ducts or a rudimentary ureter communicating with the cyst.^{4,5} Typically, seminal vesicle cysts appear as well-circumscribed retrovesical lesions demonstrating high signal intensity on T2-weighted sequences and variable signal intensity on T1-weighted sequences depending on their internal content. Increased T1 signal intensity may occur in the presence of proteinaceous material or haemorrhagic products, reflecting chronic stasis of seminal secretions.⁶ MRI also allows comprehensive evaluation of the contralateral kidney and exclusion of additional genitourinary anomalies.

The differential diagnosis of retrovesical cystic lesions includes Müllerian duct cysts, prostatic utricle cysts, ejaculatory duct cysts, vas deferens cysts, and other congenital or acquired pelvic cystic lesions. Müllerian duct cysts are typically located in the midline and are not associated with renal agenesis, while prostatic utricle cysts usually communicate with the prostatic urethra and are often detected in younger patients. Recognition of the characteristic association between a seminal vesicle cyst and ipsilateral renal agenesis is therefore fundamental for establishing the diagnosis of Zinner syndrome and distinguishing it from other retrovesical cystic entities.^{4,5}

Management depends largely on symptom severity and cyst size. Asymptomatic patients may be managed conservatively with clinical and imaging follow-up.⁷ Symptomatic patients, particularly those with significant pelvic pain, urinary symptoms or infertility, may require surgical intervention. Treatment options include transurethral resection of the ejaculatory duct, laparoscopic or robot-assisted excision of the seminal vesicle cyst, and, less commonly, open surgical approaches.⁷ Minimally invasive techniques are increasingly preferred due to lower morbidity and favourable functional outcomes. Overall prognosis is generally favourable following appropriate treatment.⁷

In the present case, multimodality imaging enabled accurate identification of the characteristic triad of Zinner syndrome, including ipsilateral renal agenesis, seminal vesicle cyst and obstruction of the ejaculatory duct, as well as the presence of a communicating ureteric remnant. Recognition of these imaging findings allowed a confident diagnosis of this rare congenital entity.

Conclusion

In summary, Zinner syndrome is a rare but important congenital anomaly of the mesonephric duct complex. Recognition of the characteristic imaging triad—renal agenesis, seminal vesicle cyst and ejaculatory duct obstruction—is crucial. MRI plays a central role in confirming the diagnosis, characterising associated anomalies, and guiding therapeutic planning.^{1,4,5}

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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