Unilateral scrotal discomfort in a patient with known renal agenesis: do not forget about Zinner syndrome

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DESCRIPTION

A 25-year-old patient was referred by his physician to our radiology department for a scrotal ultrasound (US) because of discomfort and occasional dull pain in the left hemiscrotum in the last year. He had no previous scrotal or inguinal surgery and this was his first scrotal imaging. The patient reported a history of left renal agenesis diagnosed with ultrasound and a subsequent upper abdominal MRI when he was 12.

The scrotal US scan showed a thickened and inhomogeneous, hypoechoic left epididymis (figure 1A) with normal vascularisation at Doppler imaging (figure 1B), findings consistent with ejaculatory duct obstruction. Both testicles were normal for size, echotexture and vascularisation; no significant hydrocele or varicocele was found; scrotal walls were normal. The additional use of a convex probe showed a left periprostatic cyst-like structure.

Given these findings and the anamnestic report of renal agenesis, a pelvic MRI was performed, confirming a gross ipsilateral cystic dilatation of the left seminal vesicle (figure 2). Unilateral renal agenesis, ipsilateral seminal vesicle cysts and ejaculatory duct obstruction represent a triad of mesonephric (Wolffian) duct anomalies consistent with Zinner syndrome.

This syndrome can remain asymptomatic or become symptomatic usually between the second and fourth decade of life. Symptoms are often non-specific, mostly related to difficulties in voiding and ejaculation and perineal or scrotal pain; it can also cause infertility in up to 45% of patients.[1] It is considered the male counterpart of Mayer-Rokitansky-Kuster-Hauser female syndrome.[2]

Imaging allows the correct diagnosis, which is sometimes incidental, ensuring proper differentiation with other pelvic cystic masses. Seminal vesicle cysts should be differentiated from other cysts such as prostatic utricle cyst (an area of focal dilatation that occurs within the prostatic utricle), mullerian duct cyst (a cyst that arises from remnants of the Müllerian duct) and ejaculatory duct cyst (usually intraprostatic, due to obstruction of the ejaculatory duct which in turn can either be congenital or secondary).[3]

Ultrasound is usually the first diagnostic step because it does not involve radiation exposure, it is inexpensive and noninvasive. Transabdominal US is preferred because of less discomfort for the patient, whereas transrectal US provides excellent visualisation of seminal vesicle cysts and can be a good choice for the follow-up.[4]

MRI on the other hand allows a multiplanar study, facilitates the differential diagnosis with other pelvic cystic malformation, and can be useful for surgical planning.[5] Typical findings are paramedian and periprostatic cysts, hypointense on T1-weighted and hyperintense on T2-weighted images, without contrast enhancement after gadolinium intravenous administration.[6] If the cysts have proteinaceous or haemorrhagic components, the T1-weighted signal increases.[7]

When cysts are asymptomatic or paucisymptomatic or are less than 5 cm in diameter, conservative medical therapy is indicated with follow-up through first-level examinations such as transrectal ultrasound.[8]

Surgical treatment is indicated in the case of symptomatic patients, when the cysts are more than

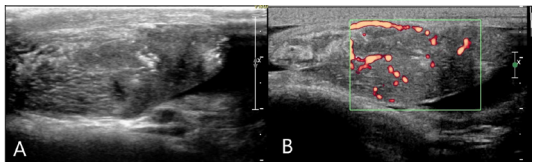

Figure 1 Ultrasound sagittal plane imaging of the body of the left epididymis, in (A) inhomogeneously hypoechoic and thickened. In (B) power Doppler imaging shows normal vascularisation, thus virtually excluding the possibility of inflammation. A small amount of anechoic fluid consistent with hydrocele surrounds the left epididymis and testicle.

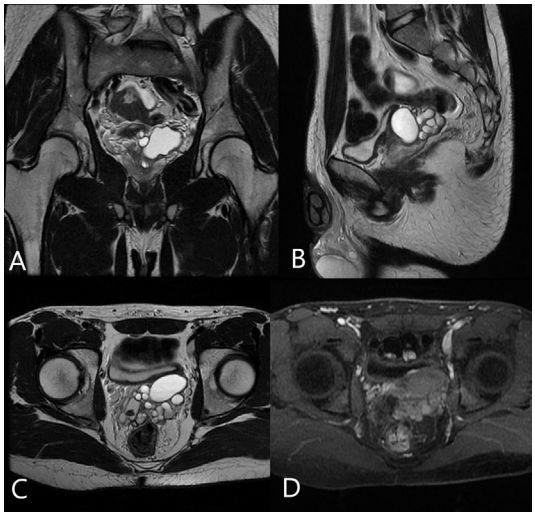

Figure 2 1,5 Tesla MRI T2-weighted pelvic images on coronal (A), sagittal (B) and axial (C) planes showing gross cyst-like dilatation of the left seminal vesicle. No abnormal vascularisation of the vesicle can be seen after IV administration of contrast media (D). No other pelvic lesions nor free fluid or enlarged lymph nodes are demonstrated.

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5 cm in diameter or in case of infertility. The main options are transurethral resection of the ejaculatory duct and laparoscopic or robotic vesiculectomy.[9]

Patient's perspective

I did not know anything about this syndrome and I could have never expected that such mild symptoms could be the beginning of a diagnostic path leading to this conclusion. I am still somewhat confused, I am young and would like to have a family and in the end knowing about Zinner syndrome could be useful to deal with my condition in the best way possible. I am also fascinated that this unusual diagnosis was possible just by using non-invasive medical imaging.

Learning points

- Zinner syndrome consists of a triad of unilateral renal agenesis, ipsilateral ejaculatory duct obstruction and seminal vesicle cysts. Its symptomatology usually begins between the second and fourth decades, with non-specific symptoms. In some cases, it may be asymptomatic and represent an incidental finding.
- The first-line examination is represented by ultrasound (US), which in association with the history and symptoms allows the diagnostic suspicion. Confirmation is obtained with MRI, which should be preferred to CT because of the absence of ionising radiation and the possibility of a more accurate anatomic pelvic study.
- Conservative treatment with periodic follow-up is reserved for asymptomatic patients or those with cysts less than 5 cm in diameter; in cases of disabling symptoms, cyst size greater than 5 cm, failure of medical therapy and infertility, surgical treatment (laparoscopic or robotic) is preferred.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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