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Abstract

Zinner syndrome is a rare congenital anomaly of the urogenital system resulting from an in utero insult during the first trimester. This entity comprises a triad of unilateral renal agenesis/dysgenesis with ipsilateral seminal vesical and ejaculatory duct obstruction. This combination of urinary and genital abnormalities occurs because of the closely related embryological origin of these structures from the distal mesonephric (Wolfian) duct. Nearly 200 cases of seminal vesical cysts with ipsilateral renal agenesis have been reported in the literature. The affected person generally presents in early adulthood when the reproductive activity commences. In this report, we present a case of a 22-year-old male with complaints of painful ejaculation and occasional backache more on the right side. There were no other complaints of lower urinary tract symptoms, trauma, or hematospermia. Past medical and surgical history was insignificant. Physical examination was unremarkable. Semen analysis revealed oligospermia with a sperm concentration of 8.5 million/mL. The radiological investigation was initiated with an ultrasound examination of the abdomen and scrotum. A large lobulated tubular cystic lesion was noted in the pelvis in retrovesical space indenting the right posterolateral aspect of the urinary bladder, measuring approximately 6 × 3 cm. There was no evidence of communication with the urinary bladder. The lesion was anechoic with no internal echoes or soft tissue components. The right seminal vesicle was not visualized. The left seminal vesicle appeared normal (►Fig. 1a, b). On the sonological examination of the upper abdomen, the right kidney was not visualized in the right renal fossa. It could not be traced elsewhere in the abdomen due to poor window from bowel gas. The left kidney was enlarged with preserved

Introduction

Zinner syndrome (ZS) consists of a triad of unilateral renal agenesis/dysgenesis, ipsilateral seminal vesical cysts, and ipsilateral ejaculatory duct obstruction resulting from the developmental arrest of the caudal mesonephric duct. This entity is one of the rarest congenital anomalies, with the frequency reported to be 0.0046%.¹ Two hundred cases of seminal vesicle cysts associated with ipsilateral renal agenesis have been reported in the literature until 2009.² The diagnosis of this entity is primarily made with the help of imaging. Magnetic resonance imaging (MRI) helps to rule out other causes of pelvic cysts with clear delineation of the anatomical structures.

Case History

A 22-year-old male patient presented to the internal medicine outpatient department with complaints of painful...
corticomedullary differentiation in its orthotopic position (► Fig. 2a, b). Bilateral testicles were normal with mildly dilated right vas deferens. Mild hydrocoele was noted in the right scrotal sac. The rest of the study was essentially normal.

After that, the patient was submitted to the MRI for further evaluation of the abnormal findings. The sonological findings were confirmed on MRI. The retrovesical cystic lesion was diagnosed as the right seminal vesical cyst, deciphered from precise anatomical detail from MRI (► Fig. 3). There was an elongated, dilated, tubular cystic lesion seen in the right paramedian region in the pelvis, extending superiorly from the seminal vesical cyst with a blind end at the level of L5-S1 vertebra. This structure was showing fluid signal intensity with T1 hypointense and T2 hyperintense signal. There was no evidence of any renal tissue noted at its proximal end (► Figs. 4 and 5). The right kidney was not visualized anywhere in the abdomen. This structure was considered a dilated remnant right ureter with ectopic drainage into the right seminal vesicle based on the anatomical location and morphology (► Figs. 6 and 4a). The left kidney was noted in the left renal fossa with compensatory hypertrophy (► Fig. 4b). These imaging features of a seminal vesical cyst, ipsilateral renal agenesis with rudimentary ureteric bud, and mild ectasia of vas deferens (probably due to obstruction) were consistent with the diagnosis of ZS, portraying the classic triad. The patient was referred to the Urology department for cyst aspiration, which was successfully performed. The patient did not show up for the follow-up visits.

Discussion

ZS is one the rarest urogenital congenital anomaly, comprising of a triad of seminal vesical cysts, ipsilateral renal agenesis, and ejaculatory duct obstruction. This entity was first identified by Zinner in 1914,3 and after that, 200 cases of seminal vesical cysts with ipsilateral renal agenesis were reported in the literature until 2009.2 The patients usually
The management includes surveillance of asymptomatic patients with transrectal sonography preferably. Symptomatic cases are treated with surgical aspiration of the seminal vesicle cysts through perineal or laparoscopic approach or percutaneous cyst drainage or a transurethral cyst deroofing to relieve the ejaculatory duct obstruction.10

Patients of ZS, a rare urogenital congenital anomaly, present with nonspecific symptoms of the lower urinary tract, and the diagnosis primarily depends on imaging studies. Two-thirds of congenital seminal vesical cysts are associated with abnormal development of other ipsilateral mesonephric duct derivatives of the urinary and genital system.2,6

Whenever seminal vesical cysts are encountered it is always better to check for associated urogenital anomalies, suspecting ZS which can be diagnosed with the triad of seminal vesical cysts, ipsilateral renal agenesis, and ejaculatory duct obstruction, of which the former two components of the triad can be diagnosed with various imaging modalities which are complementary to each other and help provide an accurate diagnosis, among which MRI is considered to be the prime modality of choice for imaging the genital tract developmental anomalies.

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Conflict of Interest
None.

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Fig. 6 Coronal 2D FIESTA fatsat MRI image of the upper abdomen showing absence of right kidney in right renal fossa (blue arrow) and compensatory hypertrophy of left kidney in left renal fossa (red arrow).