Case Report

Cystic dysplasia of the rete testis associated with ipsilateral renal agenesis: A case report

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ABSTRACT

Cystic dysplasia of the rete testis is a rare benign abnormality seen in the pediatric population. Diagnosed by scrotal ultrasonography, this lesion is often associated with congenital renal tract anomalies, most commonly ipsilateral renal agenesis or multicystic dysplastic kidney. Treatment traditionally involves orchiectomy or testicular sparing surgery; however, conservative treatment with surveillance has been used as an appropriate alternative. We present the case of a 5-year-old boy with cystic dysplasia of the rete testis with associated unilateral renal agenesis.

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Introduction

Cystic dysplasia of the rete testes is a rare benign testicular lesion of childhood characterized by irregular or tubular cystic spaces in or adjacent to the mediastinum testis [1]. While it may be an isolated anomaly, it has been shown to be associated with numerous congenital renal tract anomalies, most commonly ipsilateral renal agenesis and multicystic dysplasia of the kidney [2,3].

Case report

We present a case of a 5-year-old boy with painless left scrotal enlargement. His medical history is notable for left renal agenesis, reported on neonatal renal ultrasound at an outside facility. The patient did not report any history of scrotal pain or trauma.

Ultrasound examination of the scrotum demonstrated an asymmetrically enlarged left testis (3.4 × 1.3 × 2.4 cm) compared to the right (2.3 × 0.9 × 1.4 cm). The right testis and epididymis were unremarkable (Fig. 1). The left testicular parenchyma was significantly replaced by innumerable internal cysts and dilated tubules of the rete testis (Fig. 2A, B). The left epididymis contained multiple anechoic cysts measuring up to 1.2 cm (Fig. 2C, D).

A renal ultrasound was also performed and confirmed the absence of a left kidney (Fig. 3). The right kidney was slightly enlarged, likely related to compensatory hypertrophy.
The patient was seen by urology who opted for conservative management with yearly ultrasound surveillance.

Discussion

Cystic dilation of the rete testis is an uncommon benign testicular lesion, first described by Leissring and Oppenheimer in 1973 [4]. It is characterized by ectatic cylindrical cystic spaces in or adjacent to the mediastinum testis, usually diagnosed via scrotal ultrasonography or pathologic examination [1,4]. While more frequently seen in men over 55 years and bilaterally, referred to as cystic ectasia, its presentation in childhood, cystic dysplasia, is rare and usually unilateral, with mean age of presentation at 6.1 years [1,2]. Its association with congenital renal tract anomalies is not yet fully understood however several proposed theories postulate on mechanisms by which they may occur.

This lesion is thought to develop between the 7th and 8th week of gestation as a result of failure of anastomosis between the caudal end of the mesonephric duct – the embryologic origin of the efferent ductules, epididymis, and vas deferens – and the germinal epithelium, which will later form the rete testis [2,4]. The cephalic end of the mesonephric duct, the ureteral bud, interacts with the metanephric mesenchyme to form the kidney. Disruption of this process may lead to agenesis or dysplasia [5]. An alternative theory proposes that the rete testis undergoes early maturation and begins secreting fluid prior to canalization of the seminiferous tubules, resulting in cystic dilatation [6]. This mechanism may explain the rarely observed cases of spontaneous regression of cystic dysplasia during puberty, likely after canalization of the seminiferous tubules, allowing for drainage of the fluid [2,7].

Fig. 1 – Grayscale ultrasound of the right scrotum in the sagittal plane demonstrates a normal, homogeneous testis, and epididymis.

Fig. 2 – Grayscale ultrasound of the left scrotum in the sagittal (A, C) and transverse (B, D) planes demonstrate significant testicular replacement by innumerable internal cysts and dilated tubules of the rete testis (A, B). The epididymis contains multiple anechoic cysts measuring up to 1.2 cm (C, D).

Fig. 3 – Grayscale ultrasound of the left renal fossa in the sagittal plane reveals the absence of the left kidney.
Historically, cystic dysplasia of the rete testis was treated with orchiectomy or with enucleation surgery of the cystic component to preserve testicular function; however, recurrence was frequently observed with testicular sparing surgery [2,3]. While it is a rare lesion to occur during childhood, unilateral scrotal enlargement is not uncommon. Conservative management with serial scrotal ultrasound examinations has been considered an appropriate alternative to surgical intervention, after other causes for unilateral scrotal enlargement, such as hydrocele, testicular torsion, varicocele, hernia, or testicular neoplasm, have been excluded [5]. Diagnostic evaluation may include ultrasound, tumor marker analysis (LDH, β-HCG, AFP), and/or tissue biopsy [2,5]. Future longitudinal studies will hopefully determine the definitive treatment for these lesions; however, reports of spontaneous regression suggest watchful waiting may be an alternative to surgery [2,7].

Authorship

All authors had access and equal role in writing the manuscript.

Patient consent

Appropriate patient consent has been obtained for this case study.

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