Unilateral renal dysplasia associated with ectopic ureter opening into ipsilateral ejaculatory duct

Kinga Kowalczyk[1], Małgorzata Baka-Ostrowska[1], Karina Felberg[1], Przemysław Kluge[2], Tomasz Dzik[2]

[1]Department of Pediatric Urology, Children's Memorial Health, Warsaw, Poland
[2]Department of Pathology Children's Memorial Health Institute, Warsaw, Poland

KEY WORDS

ectopic ureter ◗ renal dysplasia ◗ epididymal cyst ◗ seminal vesicle cyst

ABSTRACT

Authors report a rare case of mesonephric duct malformation consisting of a dysplastic kidney with an ectopic megaureter opening into the ipsilateral ejaculatory duct. It presents as a recurrent epididymal cysts, progressive testicular degenerative changes and dilatation of the seminal vesicles. Nephroureterectomy with ipsilateral orchidectomy was done. Uncharacteristic symptoms, diagnostic difficulties, and findings are described.

INTRODUCTION

Improper development of the mesonephric duct can lead to malformations of the ipsilateral genital and urinary tract [1]. Genital anomalies are observed in 10-20% of patients with unilateral renal agenesis. In boys with renal agenesis the ipsilateral testicle is usually normal, but partial agenesis of the epididymis and vas deferens can be founded in 50% [2]. The roots of this correlation reach embryological development. The epididymis, vas deferens, ejaculatory duct and seminal vesicles unfold from the mesonephric duct (Wolffian duct) and the ureteral bud arises on the same duct. If the ureteral bud develops more cranially on the mesonephric duct, the ureteral orifice will open ectopically into the posterior urethra, ejaculatory duct, seminal vesicles, vas deferens or epididymis [3, 4].

The objective of this article is to analyze a rare case of recurrent epididymal cysts and degenerative changes in the testis of a child with a dysplastic kidney associated with an ectopic ureter opening into the ipsilateral ejaculatory duct.

CASE REPORT

A 3-year-old boy, previously diagnosed with weight and growth deficit, mitral insufficiency and right renal agenesis, was admitted to the outpatient clinic because of painless, progressive enlargement of the right testicle. Scrotal palpation showed enlarged, firm right testicle without inflammation. The left testicle size was normal for patient's age. Ultrasound examination revealed enlargement of the right testicle with hypoechoic center size 1.3 cm x 0.7 cm, epididymal circular cyst about 0.5 cm in diameter and a similar lesion 1.9 cm situated more cranially. The AFP and beta-HCG serum levels were normal. Routine blood and urine analysis showed no abnormalities. The testicular biopsy together with an excision of the epididymal cysts was done. Histological examination revealed edema of the fibrous epididymal tissue with cysts, dilatation of the testicular tubules and presence of Sertoli and germ cells (Fig. 1). In further follow-up the testis became large again. Repeated ultrasound examination showed a heterogenic testicular structure with a hypoechoic central lesion, fluid layer around the testicle and enlargement of epididymis with a small cyst inside. Because of normal serum levels of AFP and beta-HCG the conservative treatment was chosen. The patient presented with no symptoms, however an increase in the cysts diameter and decrease in testicular blood flow was observed. Finally, the subsequent abdominal ultrasound revealed a new anechoic structure (sized 2.3 cm x 1.4 cm x 1.0 cm) behind the bladder. In voiding cystourethrography (VCUG) no abnormalities were detected except for extravesical pressure on the right side of the bladder wall. Pelvic magnetic resonance imaging (MRI) showed cystic dilatation of the right seminal vesicle, a cystic lesion (about 1.5 cm) behind the bladder and fluid layer around the right testicle (Fig. 2). Surgical exploration was chosen. Preoperative cystoscopy showed lack of the hemitrigone on the right side with elevation of the posterior bladder wall in this region. During extravesical, retroperitoneal exploration a small kidney (1 x 1, 5 cm) was found together with an extremely dilated ureter opening into the right ejaculatory duct. Seminal vesicles were swollen. The kidney was

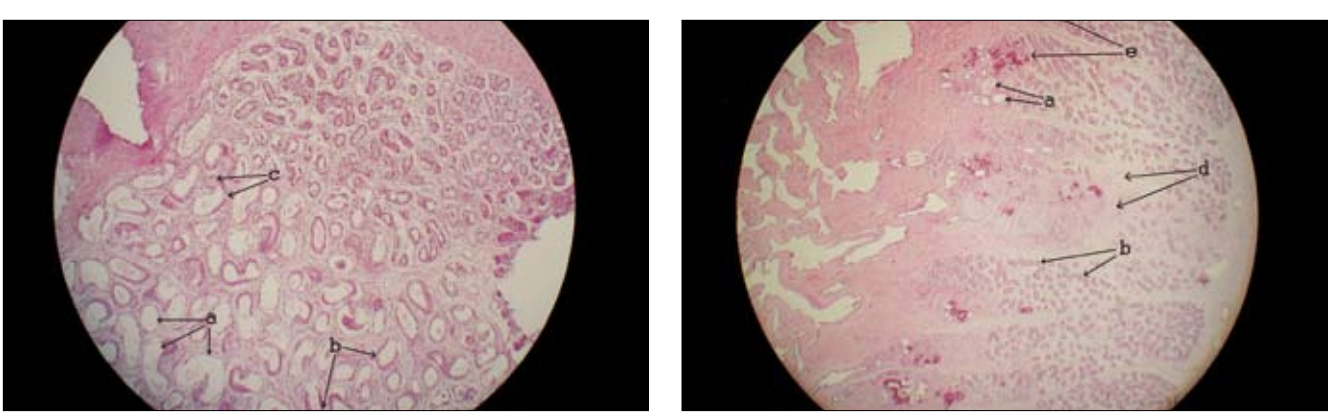

Fig. 1. Histology of the testicle: A – in the 3th year of life: a - dilatation of testicular tubules, b – presence of Sertoli cells, c – germ cells. B – in the 6th year: a - tubular dilatation, b – presence of Sertoli cells, d – advanced testicular atrophy, e - calcifications in the tubules.

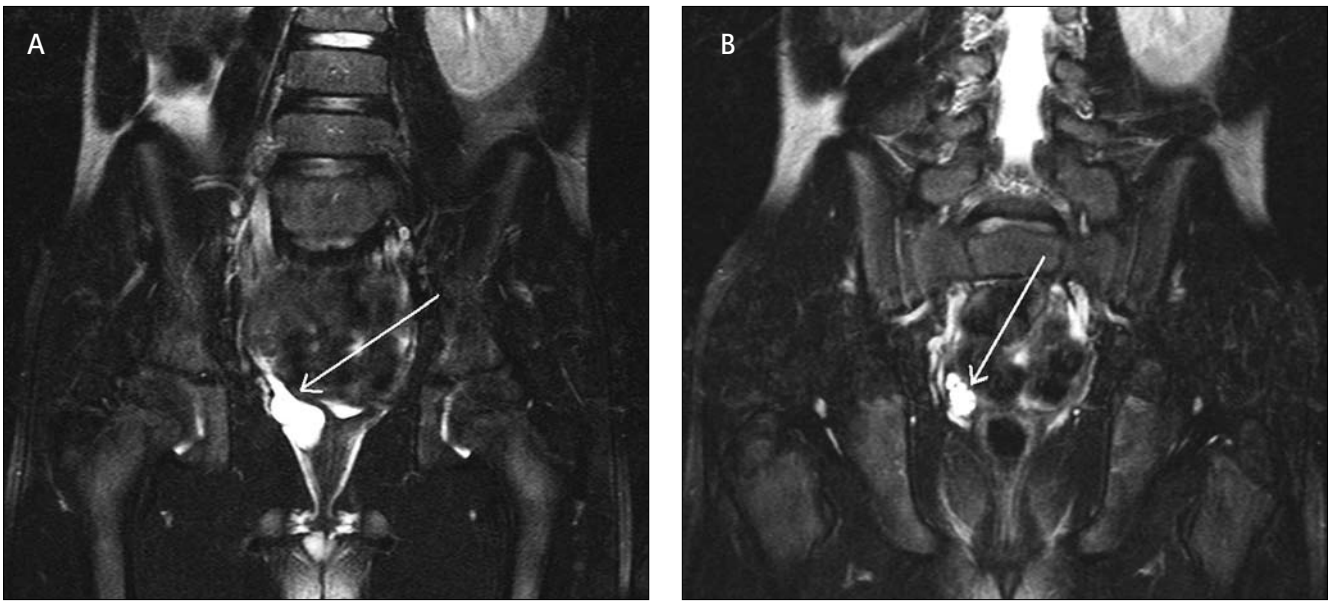

Fig. 2. Magnetic resonance imaging: A – rudimentary right megaureter. B – dilated right seminal vesicles.

removed together with the ureter and a right orchidectomy was performed. Histological examination showed a dysplastic kidney and atrophic changes in the testicle with small tubular calcifications, tubular dilatation and the presence of Sertoli and germ cells. No Leydig cells were found. There were no postoperative complications. Two years after surgery, the boy presented with no symptoms and abdominal and scrotal ultrasounds were normal.

DISCUSSION

Urine drainage into the genital tract is a very rare anomaly, but ectopic ureter opening into the epididymis is unique. Detection of this pathology is difficult because of its uncharacteristic symptoms and high risk of improper interpretation of the diagnostic images. Interesting phenomena are congenital seminal vesicles cysts which are also associated with ipsilateral renal agenesis (68-100% of cases). The ureteral remnants are found in about 13% of these patients [5]. Symptoms of an ectopic ureter that opens into the genital tract are: epididymitis, pain located in the perineum, genitalia or renal region, dysuria, frequency and urgency, but they may not appear until sexual activity [6]. In our case the first symptom was a painless testicular enlargement and epididymal cyst detected on scrotal ultrasound. The lack of the right kidney, diagnosed previously, should have focused our attention not only on the genital tract. New epididymal and extravesical cysts arose after scrotal surgery lead us to further investigation. Necessary in this case, MRI showed the presence of cysts behind the bladder and dilated seminal vesicles. But the proper diagnosis of ectopic megaureter opening into the ejaculatory duct and connected with an extremely small ectopic kidney was given by the surgeon. One year after the nephroureterectomy and orchidectomy, the child is doing well but close follow-up is needed related to infertility that may occur as described by others [1, 5, 6].

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Correspondence
Kinga Kowalczyk
Department of Pediatric Urology
Children's Memorial Health
20, Dzieci Polskich Boulevard
04-736 Warsaw, Poland
phone: +48 22 815 13 50
kingakowlaczyk@vp.pl