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CASE REPORT / *OLGU SUNUMU*

A Case Report with a Late Dıagnosis of Anterior Urethral Valves

Geç Tanı Alan Anterior Üretral Valv Olgu Sunumu

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ABSTRACT

Congenital anomalies are the leading pathologies that cause obstructive uropathy by causing stenosis in the lower urinary tract. Among these disorders, anterior urethral valves (AUV) are rarely seen and diagnosed based on the clinician's suspicion. Ultrasound findings, voiding cystourethrography (VCUG), and urethroscopy are used for diagnosis. AUV can lead to serious clinical consequences, including end-stage renal disease due to late diagnosis and treatment. In our case, unilateral kidney loss occurred due to recurrent urinary infections and nephrolithiasis. Our patient, who could not be diagnosed until the period of nephrectomy necessity, was found to have an AUV as a result of cystoscopy.

Keywords: Anterior urethral valves, obstructive uropathy, prognosis

ÖZ

Alt üriner sistemde darlığa yol açarak obstrüktif üropatiye neden olan patolojilerin başında konjenital anomaliler gelmektedir. Bu bozukluklardan anterior üretral valv (AUV), nadiren görülmekte ve klinisyenin şüphesi doğrultusunda tanı almaktadır. Tanıda ultrason bulguları, işeme sistoüretrografisi (VSUG), üretroskopi kullanılmaktadır. AUV geç tanı ve tedavi nedeniyle son dönem böbrek yetmezliği de dahil olmak üzere ciddi klinik sonuçlara yol açabilmektedir. Bu makalede anlatılan vakamızda tekrarlayan üriner enfeksiyonlar ve nefrolitiasiz nedeniyle tek taraflı böbrek kaybı meydana gelmiştir. Yapılan tanısal sistoskopi sonucu anterior üretral valv tanısı alan ve darlığı açılan hasta iyi klinik sonuçlar ile takip edilmektedir.

Anahtar Kelimeler: Anterior Üretral Valv, Obstrüktif üropati, Prognoz

INTRODUCTION

Anomalies such as posterior urethral valves, anterior urethral valves, diverticulum, and urethral atresia, cause stenosis in the lower urinary tract which leads to uropathy. Among these, anterior urethral valves are less common. It is usually diagnosed in the neonatal period (1). AUV, the etiology of which is not fully known, has been noted in the bulbous (40%); penile urethra (30%); and the penoscrotal junction (30%) (2). Urinary tract infection, which recurs by causing stenosis in the lower urinary tract, is involved in the etiology of many clinical conditions, including voiding dysfunction, incontinence, enuresis, hydroureteronephrosis, and end-stage renal failure (3). In the diagnosis, mainly voiding cystourethrography is used as well as urethroscopy, renal ultrasound, and transpenile ultrasound. Although the treatment changes according to the degree of the disease, patients are usually treated with methods such as endoscopic valve resection and urethrotomy (4).

CASE

A fifteen-year-old male patient, who was followed up for recurrent kidney stones and nephrectomy, appeared with the complaint of inability to urinate for the last 2-3 days. It was learned that in the patient's medical history, there was a cleft lip and palate repair surgery at the age of 1. The patient had recurrent febrile urinary tract infections, there were failed attempts to break a stone in the left kidney with ESWL twice when he was 7-8 years old, and a stent was attempted to be inserted due to left hydronephrosis and renal calculus, but it also failed. The left kidney was evaluated as non-functional as the result of scintigraphy and a left simple nephrectomy was performed when the patient was 9 years old. The patient, whose pathology result had revealed xanthogranulomatous pyelonephritis, had been followed up with urinary ultrasound. The patient had macroscopic hematuria when he presented to our department. His laboratory test results were as follows: Albumin/creatinine in spot urine: 147.7 mg/g (N: 0-30 mg/g), protein/creatinine: 282.5 mg/g (N:0-200 mg/g). His

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biochemical results were as follows: Urea: 47 mg/dl (N: 17-43 mg/dl), creatinine: 1.1 mg/dl (N: 0.26-0.77 mg/dl), glomerular filtration rate: 70 ml/min/1.73 m², Na: 137 mmol/L (N: 135-145 mmol) /L), K: 4.6 mmol/L (N: 3.4-5.1mmol/L), folate: 6.1 µg/L (N: 3.1-20.5 µ/L), vitamin B 12 : 196ng/L (N: 187-883 ng/L), parathormone: 126.7 ng/L (N: 18.5-88 ng/L). On USG, compensatory hypertrophy was observed in the right kidney and the post-void residual (PMR) was measured as 100 cc. A catheter was inserted in the patient due to the complaint of difficulty in urination for the last 1 week. Voiding cystourethrography was planned. After catheter insertion, urine output occurred, and cystoscopy was performed by the pediatric urologist.

A bird's eye stenosis at the 12 o'clock position in the bulbar urethra and a false area proximal to the stenosis were observed, and stenosis was treated with a cold incision. In the control examinations of the patient whose complaints regressed after the operation, albumin/creatinine in spot urine was 66.6 mg/g (N: 0-30 mg/g), while the biochemical results were as follows: Na: 141 mmol/L (N: 135-145 mmol/L), K: 5 mmol/L (N: 3.4-5.1 mmol/L), phosphorus: 3.8 mg/dl (N: 4-7 mg/dl), urea: 28 mg/dl (N: 17-43 mg/dl), creatinine: 0.78 mg/dl (N: 0.26-0.77 mg/dl), parathormone: 43.1 ng/L (N: 18.5-88 ng/L) and glomerular filtration rate: 99 ml/min/1.73 m². On USG, PMR was measured as 40 cc. The follow-up of the patient continues.

DISCUSSION

AUV is a rare cause of lower urinary tract obstructions. It has been reported that the frequency of AUV, which can also be found with anterior urethral diverticulum (AUD), is 10-30 times less than PUV(5). Suspicion plays an important role in their detection, and patients may experience renal losses when the diagnosis cannot be made (3).

Although they are thought to occur due to reasons such as incomplete urethral duplication and congenital cystic dilatation of the urethral diverticula in the periurethral glands in the embryological period, the etiology is still not clear (6). AUV, which can be located anywhere, are most commonly seen in the bulb, penile urethra, and penoscrotal junction, and it has been reported that they can also be found in the navicular fossa in a few cases (2,4,7). In our patient, the stenosis was located in the bulbous.

The clinical findings of AUV vary according to the age of the patient and the degree of obstruction. While the majority of cases are detected in the neonatal period, they can rarely be diagnosed in late childhood as in our patient. Symptoms of urinary infection such as fever, vomiting, dehydration, and septicemia are observed. In children and adolescents, findings such as voiding difficulty, weak urine flow, urinary retention, recurrent urinary tract infections, dysuria, and hematuria are prominent (4,5). Our patient has been followed up since he was 7-8 years old due to the persistence of complaints such as decreased urine output, hematuria, frequently recurring urinary tract infection, and nephrolithiasis, and unilateral nephrectomy was needed due to recurrent stones and scars.

According to the classification developed by Firlit et al., there are four types of AUV. Type 1 is the mildest form, and there is minimal enlargement in the proximal urethra, whereas hydroureteronephrosis, high-grade VUR, enlargement of the bladder, and pelvicalliectasis are seen in type 4 obstructions (8). Our patient was evaluated as type 4 because of renal loss. However, the delayed diagnosis was also considered an important factor aggravating the prognosis.

As in all other obstructive disorders, voiding cystourethrography is the most valuable method in diagnosis. It is performed with the patient in the anterior oblique position in children, and provides information about the location and degree of stenosis (4,5,7,9). Apart from VCUG, retrograde urethrography, intravenous urography, nuclear imaging, ultrasound, urethroscopy, and cystoscopy are other diagnostic methods (9). Ultrasound provides information about bladder dilatation, trabeculation, urethral dilatation, renal parenchymal, and pelvic changes. It is stated that transpenile ultrasound performed during micturition may also be useful in diagnosis (4). In our case, grade 1 echo increase in the right renal parenchyma, enlarged kidney size, and grade 1 hydronephrosis were detected, while the bladder was evaluated as normal. However, he had no history of VCUG or cystoscopy taken until the time he presented to our department.

Although it varies according to the patient, open and closed methods are used in the treatment. Valve excision with urethrotomy and segmental urethrotomy is preferred in severe urethral anomalies, and urinary extravasation, fistula, and stenosis may occur as complications. Endoscopic valve resection is the most preferred method in all age groups, and can lead to urethral dilatation, stenosis, and fistula (10). Diagnostic cystoscopy was performed on our patient, who could not have VCUG performed, and the detected stenosis was successfully treated with a cold incision.

Evaluated in line with clinical suspicion, when the diagnosis of AUV is delayed until the adolescent-adult age, it can cause serious urinary problems (4,10). In a study conducted in 2010, complete recovery was achieved in 108 of 139 patients who were examined retrospectively, however, it was observed that renal functions did not improve in 31 patients (1). As reported by Aygün et al., AUV was detected in two patients with end-stage renal disease who were examined before transplantation, and stenosis was treated in one patient while reconstruction was planned for other patients due to serious changes (3). In these patients, stasis and recurrent infections due to stenosis also predispose them to stone formation. Çetin et al. reported that stone formation due to stasis occurred in a pediatric case with both AUV and AUD (7). Similarly, in our case, the diagnosis of AUV was delayed and it was diagnosed after kidney loss due to recurrent stones.

CONCLUSION

AUV is a rare obstructive uropathy. Therefore, there are delays in diagnosis, and renal prognosis worsens. Irreversible urinary system disorders are seen in patients who have reached adulthood at the time of diagnosis. Preventing renal losses

with rapid diagnosis and treatment seems possible with an increase in the awareness of physicians on the issue. It should be a diagnosis that must be kept in mind, especially in patients with a history of recurrent urinary infections and stones.

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REFERENCES

1. Routh JC, McGee SM, Ashley RA, Reinberg Y, Vandersteen DR. Predicting renal outcomes in children with anterior urethral valves: a systematic review. J Urol 2010;184(4):1615-9. doi:10.1016/j.juro.2010.03.119
2. Scherz HC, Kaplan GW, Packer MG. Anterior urethral valves in the fossa navicularis in children. J Urol 1987;138(5):1211-3. doi:10.1016/s0022-5347(17)43552-x
3. Aygün C, Güven O, Tekin MI, Peşkircioğlu L, Ozkardeş H. Anterior urethral valve as a cause of end-stage renal disease. Int J Urol 2001;8(3):141-143. doi:10.1046/j.1442-2042.2001.00270.x
4. Kibar Y, Coban H, Irkilata HC, Erdemir F, Seckin B, Dayanc M. Anterior urethral valves: an uncommon cause of obstructive uropathy in children. J Pediatr Urol 2007;3(5):350-3. doi:10.1016/j.jpurol.2007.01.197
5. Paulhac P, Fourcade L, Lesaux N, Alain JL, Colombeau P. Anterior urethral valves and diverticula. BJU Int 2003;92(5):506-9. doi:10.1046/j.1464-410x.2003.04380.x
6. Karnak I, Senocak ME, Büyükpamukçu N, Hiçsönmez A. Rare congenital abnormalities of the anterior urethra. Pediatr Surg Int 1997;12(5-6):407-9. doi:10.1007/BF01076951
7. Çetin S, Koparal MY, Bulut EC, Gürocak S, Tan Ö. Anterior urethral valve and anterior urethral diverticulum with an intradiverticular stone: A case report and a literature review. J Urol Surg 2018;5:109-11. doi:10.4274/jus.1420
8. Firlit RS, Firlit CF, King LR. Obstructing anterior urethral valves in children. J Urol 1978;119(6):819-21. doi:10.1016/s0022-5347(17)57642-9
9. Jana M, Gupta AK, Prasad KR, Goel S, Tambade VD, Sinha U. Pictorial essay: Congenital anomalies of male urethra in children. Indian J Radiol Imaging 2011;21(1):38-45. doi:10.4103/0971-3026.76053
10. Zia-ul-Miraj M. Anterior urethral valves: a rare cause of infravesical obstruction in children. J Pediatr Surg 2000;35(4):556-8. doi:10.1053/jpsu.2000.0350556