Unusual Surgical Treatment Option for an Uncommon Disease: Ureterocutaneostomy in an Emphysematous Pyelonephritis Patient

Nadir Bir Hastalık İçin Olağandışı Cerrahi Tedavi Seçeneği: Amfizematöz Piyelonefritli Hastada Üreterokutanostomi

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ABSTRACT

Emphysematous pyelonephritis (EPN) is severe necrotising inflammation of renal parenchyma. Because of the devastating consequences of the pathology aggressive treatment can prevent permanent renal damage. Fifteen-year-old-girl patient without medical follow-up applied to emergency room with fever and vomiting. Computed Tomography (CT) revealed EPN. After diagnostic procedure minimal invasive procedures like double J catheter and nephrostomy tube insertion failed. While taking into account family's treatment and medical follow-up compliance, also severe dilatation in both ureters made bilateral ureterocutaneostomy (UC) to be considered as a feasible option. UC creation can be thought as a safe and effective alternative surgical procedure. Pediatric neurogenic bladder (NB) population incompatible to treatment and follow-up are candidates for EPN development. We also believe that UC can be beneficial in terms of acute and long-term management of EPN in NB patients who the clinician thinks will not be compatible with complex surgical intervention, follow-up and treatment.

Key Words: Children, Emphysematous Pyelonephritis, Neurogenic Bladder

ÖZ

Amfizematöz piyelonefrit (AP), renal parankimin ileri derece nekrotizan inflamasyonudur. Patolojinin yıkıcı sonuçları nedeniyle agresif tedavi kalıcı böbrek hasarını önleyebilir. On beş yaşında takipsiz kız hasta acil servise ateş ve kusma şikayeti ile başvurdu. Bilgisayarlı Tomografi (BT) AP şeklinde rapor edildi. Tanı sürecinden sonra bilateral double J kateter veya nefrostomi gibi minimal invaziv girişimler başarısız oldu. Hastanın postür bozukluğu nedeniyle nefrostomi tüpü ile drenaj yapılamadı. Ailenin tedavisi ve tıbbi takip uyumu dikkate alınarak, her iki üreterde de ciddi dilate olması bilateral üreterokütanostominin (ÜK) uygun bir seçenek olarak düşünülmesine neden oldu. ÜK oluşturulması güvenli ve etkili alternatif bir cerrahi prosedür olarak düşünülebilir. Tedavi ve takibe uyumsuz pediatrik nörojen mesane (NB) popülasyonu, AP gelişimi için adaydır. Klinisyenin karmaşık cerrahi müdahale, takip ve tedavi ile uyumlu olmayacağını düşündüğü NB hastalarında ÜK'nin AP'in akut ve uzun süreli yönetimi açısından faydalı olabileceğine inanıyoruz.

Anahtar Kelimeler: Çocuk, Amfizematöz, Piyelonefrit, Nörojen Mesane

INTRODUCTION

EPN is rare and aggressively progressive urinary tract infection characterized by gas collection within renal parenchyma, collecting system or perirenal tissue (1). The first case of EPN in adults was reported in 1898, and the first case of EPN in a pediatric patient was reported in 1985 (2,3). EPN is extremely rare in pediatric population (1). EPN is extremely rare in the pediatric population, to date only eight pediatric patients have been reported in the literature, to our knowledge) (1,3,4-9). Most of the cases are documented from adult population (10). Aggressive treatment is mandatory because of the high mortality rate (12-50%) (11). Neurogenic bladder (NB), obstructive pathologies of urinary tract (ureterocele,

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ectopic ureter, ureteropelvic junction obstruction), acquired immunodeficiency, renal stones or previous history of nephro-urological malformation surgery are reported causes of EPN pediatric population (1,4-9). Treatment could be medical alone or a combination of medical and surgical interventions.

CASE REPORT

Fifteen-year-old-girl patient without medical follow-up applied to emergency room with fever and vomiting. Patient had a history of meningocele operation without shunt. Serious postural disorder secondary to severe scoliosis, club foot deformity and immobility of lower extremity detected in clinical examination of the patient. Abdominal examination was insignificant because of the patient's mental status. Laboratory analyses was as follows: WBC: 30.6x103/mm^3, HGB:7 g/dl, PLT: 4696x103/mm^3, BUN:59 mg/dL, creatinine: 2.17 mg/dL, Na-122 mmol/L, K-4.36 mmol/L, CRP-367 mg/L Antibiotics and intravenous fluid resuscitation for electrolyte imbalance started and Foley catheter inserted and purulent drainage of urine observed.

Urinary ultrasound in emergency room revealed parenchymal hyperechogenicity in both kidneys, severe dilatation in pelvicalyceal system – anteroposterior diameter of right and left kidney reported to be 37mm and 45 mm, respectively. Also, severe thickness and mucosal irregularity of bladder wall reported. Because of the inability of patients to express herself and insignificant clinical examination findings abdominal computerized tomography (CT) was planned. Enlarged kidneys, intraparenchymal and intracalyceal gas with an air fluid level confirmed diagnosis EPN as a cause of acute renal failure (Figure 1). Bilateral severe ureteral dilatation and very thick-walled fibrotic bladder detected in CT. *E. coli* isolated from the urine samples of the patient.

After diagnostic procedure bilateral double J catheter insertion for upper urinary tract drainage was planned. Because of the severe trabeculation of mucosa, fibrotic appearance of bladder that compromised ureteral orifices to be detected, procedure failed. Drainage with nephrostomy tube was impossible because of the postural disorder of the patient. While taking into account family's treatment and medical follow-up compliance, also severe dilatation in both ureters made bilateral led us to consider UC as a feasible option. After discussion with the family operation was completed. On post-operative fifth day, clear urine drainage obtained from UC and blood analyses was completely normal. She discharged from the hospital on postoperative first week. The option of bladder augmentation will be offered to family after full recovery.

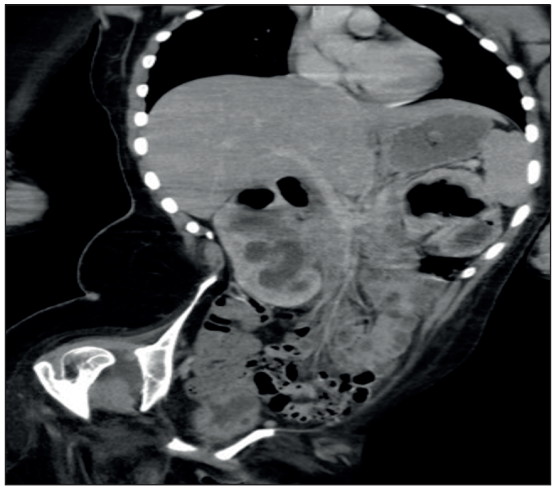

Figure 1: Emphysemateus change within renal parenchyma.

DISCUSSION

EPN is rare and aggressively progressive urinary tract infection characterized by gas collection within renal parenchyma, collecting system or perirenal tissue (1). The first case of EPN in adults was reported in 1898, and the first case of EPN in a pediatric patient was reported in 1985 (2,3). EPN is extremely rare in the pediatric population, to date only eight pediatric patients have been reported in the literature, to our knowledge (1,3). Most of the cases are documented from adult population (4). Aggressive treatment is mandatory because of the high mortality rate (12-50%) (5). Neurogenic bladder (NB), obstructive pathologies of urinary tract (ureterocele, ectopic ureter, ureteropelvic junction obstruction), acquired immunodeficiency, renal stones or previous history of nephro-urological malformation surgery are reported causes of EPN pediatric population (1,4-9). Unlike adults, high renal glucose levels secondary to diabetes mellitus (DM) is not detected to be a comorbidity factor in pediatric population (1). There is only one reported case without apparent health issue from Nigeria (6). Comorbidity of the presented case was bilateral functional obstruction due to severe NB.

E. coli is the most common EPN causing pathogen (70%). Especially in patients with DM and urinary tract obstruction *E. coli* and *K. pneumonia, Proteus mirabilis, Klebsiella pneumoniae*, *group D Streptococcus, coagulase-negative Staphylococcus*, and *Enterobacteriaceae* are other causative pathogens (7). Rare organisms such as coagulase negative *Staphylococcus*, *Clostridium, Candida species* and *Aspergillus fumigates* have also been reported (8,9). Rare pathogens like *A. schaalii, Actinomyces turicensis, Prevotella bergensis*, and *Prevotella disiens* isolated from the urine sample of the case reported by Kitano et al. (1) *E. coli* in concordance with the literature isolated from the urine culture of the presented case. There are two speculated mechanisms for EPN development. Presence of gas-producing pathogen in addition to local tissue defect encourage tissue destruction and inhibition of locally produced gas thought to be cause of EPN. Other speculated mechanisms are that the increased levels of glucose in the tissues together with decreased blood supply to the kidneys contributes to the anaerobic metabolism of glucose and lactate

by the organisms and thereafter the production of gases like carbon dioxide, hydrogen, nitrogen, oxygen and methane by the gas-forming organisms (10). CT is a recommended radiological tool for classification of EPN, which was developed by Huang and Seng (8). According to the CT findings, the presented case classified as class 2, which is defined as gas in the renal parenchyma without extension to the extrarenal space.

EPN is a life-threatening condition with 12-50% of mortality (5). There is no consensus regarding treatment options in pediatric population also.

Treatment options for EPN are (9):

1-Fluid and electrolyte replacement, correction of acid-base imbalance, 2-Antibiotic treatment, 3-Percutaneouus drainage (PCD), 4-Urgent nephrectomy or subsequent surgical intervention. Especially in a pediatric population conservative treatment should be promptly attempted for prevention of fulminant course and nephrectomy. Treatment options can be combined according to the patient's clinical course. In this case after administration of subsequent intravenous fluid and meropenem initiation (1gr/every 8 hours) surgical intervention was performed. For localized EPN (class 1 or 2) PCD and/or relief of the urinary tract obstruction with antibiotic combination reported to provide good outcome (8). In presented case, bilateral UC was created instead of PCD or other minimal invasive procedures because of the inconvenient bladder and postural appearance of patient. However, unsuccessful PCD, fulminant course of disease or extensive EPN may require nephrectomy (8). As a result of the literature review about EPN in pediatric population, it can be claimed that this is the first case of EPN managed with urinary diversion.

CONCLUSION

Considering kidney's irreplaceable role in growth and development of the pediatric population, UC creation can be thought as a safe and effective alternative surgical procedure. Pediatric NB population incompatible with treatment and follow-up are candidates for EPN development. We also believe that UC can be beneficial in terms of acute and long-term management of EPN in NB patients who the clinician thinks will not be compatible with complex surgical intervention, follow-up and treatment.

A consent form for the use of medical information was signed by patient's parents.

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