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Akut Poststreptokokal Glomerülonefrit ile Fındıkkıran Sendromunun Birlikteliği; Bir Olgu Sunumu

Coexistence of Acute Poststreptococcal Glomerulonephritis with Nutcracker Syndrome: A Case Report

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ÖZET

Akut poststreptokokal glomerülonefrit (APSGN), sık karşılaşılan ve tam iyileşmenin sağlanabildiği, kendi kendini sınırlayan bir hastalıktır. Ancak eşlik eden anomaliler hastalığın seyrini zorlaştırabilir. Fındıkkıran sendromu (nutcracker sendromu,NCS), sol renal venin aort ile superior mezenterik arter arasında sıkışmasıdır ve hematüri, proteinüri, yan ve karın ağrısı gibi çeşitli semptomlarla kendini gösterir. Burada gros hematüri, ateş ve periorbital ödem şikayetleri ile birlikte APSGN ve NCS tanısı alan 12 yaşında bir erkek çocuk sunuldu. Hastamızda olduğu gibi hematüriye neden olan bazı hastalıkların birlikteliğinin tanıyı zorlaştırabileceği ve prognozu kötüleştirebileceği akılda tutulmalıdır.

Anahtar Kelimeler: Akut poststreptokokal glomerülonefrit, fındıkkıran fenomeni, fındıkkıran sendromu

ABSTRACT

Acute poststreptococcal glomerulonephritis (APSGN) is a common and self-limiting condition for which complete recovery can be achieved. However, accompanying anomalies can complicate the disease prognosis. Nutcracker syndrome (NCS) is the entrapment of the left renal vein between the aorta and the superior mesenteric artery and presents with a variety of symptoms, such as hematuria, proteinuria, flank, and abdominal pain. Herein, we report a 12-year-old boy with complaints of gross hematuria, fever, and periorbital edema who was diagnosed with concurrent APSGN and NCS. It should be kept in mind that the coexistence of some diseases that cause hematuria, as in our patient, may complicate the diagnosis and worsen the prognosis.

Keywords: Acute poststreptococcal glomerulonephritis, nutcracker phenomenon, nutcracker syndrome

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INTRODUCTION

Acute poststreptococcal glomerulonephritis (APSGN) is the most seen acute glomerulonephritis in children, characterized by macroscopic hematuria, hypertension, and decreased levels of complement C3 (1). APSGN is an immune complex-mediated glomerular disease triggered by pharyngitis or skin infections due to group A β-hemolytic streptococcus (GAS) (2).

The nutcracker phenomenon (NCP) occurs when the left renal vein (LRV) is compressed between the aorta and the superior mesenteric artery (SMA) (3). The nutcracker syndrome (NCS) is defined as NCP accompanied by symptoms and signs, such as hematuria, proteinuria, and left flank pain (4). The prevalence of NCS is not fully known because definitive diagnostic criteria for NCS have not yet been established, and it is difficult to diagnose due to the variety of non-specific symptoms (3,4).

In this paper, we report on a 12-year-old boy diagnosed with concurrent APSGN and NCS. To our knowledge, this is the first reported case of the coexistence of APSGN and NCS.

CASE REPORT

A 12-year-old boy was admitted to our hospital with complaints of gross hematuria, fever, fatigue, nausea, intermittent periorbital edema, and abdominal distension lasting for 10 days. He also had a history of a sore throat two weeks earlier and intermittent left-sided flank pain.

The examination of the patient revealed that he had gained about 4 kilograms within the past month. He was hypertensive with a blood pressure (BP) of 140/93 mmHg (95th percentile for his height – 124/78 mmHg). He had periorbital and pretibial 2+ edema, as well as ascites.

The patient's initial laboratory findings were as follows: hemoglobin, 12.4 g/dL; hematocrit, 39%; white blood cell count, 9 270/mm3; platelet count, 473 000/mm3; urea, 28 mg/dL; serum creatinine, 0.57 mg/dL; uric acid, 4.4 mg/dL; sodium, 140 mmol/L; potassium, 5.4 mmol/L; calcium, 9.4 mg/dL; phosphorous, 4.8 mg/dL; total protein, 62 g/L; albumin, 3.3 g/dL; triglyceride, 235 mg/dL; total cholesterol, 248 mg/dL; 24-hour urine protein, 56 mg/m2/h; anti-streptolysin O antibody, 1,384 IU/L; C3, 0.23 g/L (normal values: 0.6–1.5 g/L); and C4 0.37 g/L (n: 0.09–0.4 g/L). Urinalysis findings were as follows: pH, 6; density, 1,014, erythrocyte, 3+; protein, 2+; leukocytes, 3+. In the urinary microscopic examination, erythrocytes were abundant, and the leukocyte count was 10–15 cells per high-power field. Urinary Doppler ultrasonography (DUS) findings were normal, but urinary DUS showed that the left renal vein (LRV) running between the aorta and the SMA was very narrow, the anteroposterior diameter of LRV was 8 mm, and the pre-compression to compression ratio of the LRV was 3.3, consistent with NCP. On renal USG, LRV entrapment was observed, which was caused by the temporary swelling of the kidneys due to APSGN; therefore, urinary DUS was repeated, and the findings were consistent with NCP. We started salt and fluid restrictions and administered ampicillin for GAS to prevent any possible outbreak in the community. An oral calcium antagonist and intravenous furosemide were administered as appropriate. The patient's creatinine level increased to 0.95 mg/dL on the third day and decreased to normal on the sixth day. Proteinuria that was in the nephrotic range regressed to the nephritic range, and diuresis resolved within one week. On the 10th day, his C3 level increased to 0.82 g/L. Macroscopic hematuria and nephritic-range proteinuria resolved in two weeks and two months, respectively. However, microscopic 1+ hematuria persisted for the first year after hospitalization. The patient's history of intermittent mild left-sided flank pain was associated with NCS. When his medical records were examined in terms of NCS, microscopic hematuria was 1+, and the red blood cell count was 8-10 intermittently.

DISCUSSION

APSGN is the major cause of acute glomerulonephritis among children, especially in low-income countries and mostly caused by GAS. Herein, we report a patient with APSGN diagnosed based on hypertension, edema, and oliguria with macroscopic hematuria that developed two weeks after high ASO titers during an episode of pharyngitis and concurrent NCS. Streptococcal cultures are reported to be positive in 10-70% of patients (1,2). We did not identify GAS in bacteriological culture.

APSGN is a self-limiting benign disease with a low mortality rate. However, the other disease that concurrently occurs with APSGN may vary the prognosis of the disease. There are a few reports in the literature describing the coexistence of APSGN with congenital anomalies of the kidney and urinary tract (CAKUT), Henoch-Schönlein purpura, and immunoglobulin A nephropathy (IgAN) (5). Tasic et al. reported that 2.6% of their patients with APSGN had CAKUT (5). Patients with a solitary kidney and bilateral renal impairment have been shown to have a worse prognosis (5,6).

To the best of our knowledge, this is the first report in the literature concerning the coexistence of APSGN with NCS, a rare vascular entrapment disorder that presents with a variety of symptoms, such as hematuria, proteinuria, flank and abdominal pain, varicocele, dysmenorrhea, and fatigue (3,4). The diagnosis of NCS is challenging due to the presence of various non-specific clinical criteria and the existence of asymptomatic patients with a normal variant of dilatation of the LRV (4). A pre-compression to compression ratio of LRV over 2.25 has been reported to have more than 91% specificity and sensitivity for NCS (7). In this case report, the patient's history of intermittent microscopic hematuria, left flank pain,

and urinary DUS findings were consistent with NCS. The diagnosis of NCS was made incidentally during an episode of APSGN. The prognosis of the patient was excellent; however, microscopic hematuria and flank pain persisted intermittently over the one-year follow-up following the APSGN episode. Whether the ongoing microscopic hematuria was due to NCS or a long-term complication of APSGN needs further evaluation based on the long-term follow-up data of the patient. The antibodies and complement proteins activated during GAS infection in APSGN causes deposits in glomeruli (1,2). The local deposition of the immune complex in patients with APSGN can cause compression and congestion of the renal venous system and lead to NCS or the exacerbation of pre-existing NCS. Since there was no imaging before the APSGN episode in our patient, we cannot comment on whether NCS developed due to ASPGN.

The literature includes case reports on the coexistence of some diseases with NCS (8,9). Imai et al. reported a rate of 6.8% prevalence for LRV entrapment in IgAN, but they were not able to demonstrate a causal relationship between NCS and IgAN (8). In another study, it was suggested that due to the lack of information on NCP in the past, some NCS cases with hematuria were misdiagnosed with glomerular nephritis and treated with immunosuppressive agents (9). However, there is no report in the literature referring to the coexistence of APSGN and NCS. The combination of these two diseases may increase the complexity of the diagnosis and disease management.

CONCLUSION

Hematuria is one of the main presenting symptoms of kidney disease in children. The coexistence of some diseases with APSGN that cause hematuria, as in our patient, can complicate the diagnosis and worsen the prognosis. These patients must be carefully examined for etiological causes.

Patient consent:

Informed consent was obtained from the patient and his legal guardian.

REFERENCES

1. Ong LT. Management and outcomes of acute post-streptococcal glomerulonephritis in children. World J Nephrol. 2022;11:139-45.
2. Brant Pinheiro SV, de Freitas VB, de Castro GV, Rufino Madeiro BC, de Araújo SA, Silva Ribeiro TF, et al. Acute Post-Streptococcal Glomerulonephritis in Children: A Comprehensive Review. Curr Med Chem. 2022;29:5543-59.
3. Meyer J, Rother U, Stehr M, Meyer A. Nutcracker syndrome in children: Appearance, diagnostics, and treatment - A systematic review. J Pediatr Surg. 2022;57:716-22.
4. Gulleroglu K, Gulleroglu B, Baskin E. Nutcracker syndrome. World J Nephrol. 2014;3:277-81.
5. Tasic V, Ristoska-Bojkovska N, Gucev Z, Lozanovski VJ. Poststreptococcal glomerulonephritis in children with congenital anomalies of the kidney and urinary tract. Ren Fail. 2015;37:1440-3.
6. Fujinaga S, Hirano D, Nishizaki N, Kanai H, Ohtomo Y, Kaneko K, et al. Unfavorable outcome in a child with megaureter-megacystis syndrome complicated by mild acute poststreptococcal glomerulonephritis. Pediatr Int. 2010;52:895–6.
7. Hangge PT, Gupta N, Khurana A, Quencer KB, Albadawi H, Alzubaidi SJ, et al. Degree of Left Renal Vein Compression Predicts Nutcracker Syndrome. J Clin Med. 2018;7:107.
8. Imai N, Shirai S, Shibagaki Y, Kimura K. Nutcracker phenomenon in IgA nephropathy. Clin Kidney J. 2014;7:325–6.
9. Ma Z, Liu X, Ning Y, Shao J, Liu W, He X. Nutcracker phenomenon in combination with glomerular nephritis in isolated hematuria patients. Int Urol Nephrol.