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Systemic Lupus Erythematosus Presenting with Renal Vein Thrombosis: A Case Report

Renal Ven Trombozu ile Prezente Olan Sistemik Lupus Eritematozus: Olgu Sunumu

ABSTRACT

The occurrence of acute renal-vein thrombosis (RVT) is a possible but rarely described complication of systemic lupus erythematosus (SLE). It has not been reported to occur as the initial presentation in lupus patients. We hereby describe a 17-year-old female with renal vein thrombosis due to SLE on initial presentation. The patient presented with flank pain, flank tenderness and fever. She was treated with anticoagulation, the mainstay of therapy for RVT in general. With appropriate diagnosis and anticoagulation therapy, our patient had a benign course during 6 months of follow-up.

KEY WORDS: Systemic lupus erythematosus, Renal vein thrombosis

ÖZ

Akut renal ven trombozu (RVT) sistemik lupus eritematozus (SLE) seyrinde görülen nadir bir komplikasyonudur. Lupus hastalarında ilk bulgu olarak RVT görülmesi literatürde daha önce bildirilmemiştir. Bu yazıda ateş, yan ağrısı ve hassasiyeti ile başvuran, eş zamanlı RVT ve SLE tanısı alan hasta sunulmuştur. Hasta antikoagulasyon ile 6 aydır takip edilmektedir.

ANAHTAR SÖZCÜKLER: Sistemik lupus eritematozus, Renal ven trombozu

Ezgi ÇOŞKUN YENİGÜN
Fatih DEDE
Serhan Vahit PİŞKİNPAŞA
Ramazan ÖZTÜRK
Nihal ÖZKAYAR
Didem TURGUT

Numune Training and Research Hospital,
Department of Nephrology,
Ankara, Turkey

INTRODUCTION

Patients with systemic lupus erythematosus (SLE) are prone to thrombotic complications. Thrombotic thrombocytopenic purpura like syndrome, anticardiolipin syndrome and renal vein thrombosis are well-documented vascular complications of SLE (1). Renal vascular complications are not infrequently encountered in SLE. Glomerlar pathology is considered the most important in the lupus nephritis, and vascular complications are forgotten by clinicians. We present a case with renal vein thrombosis (RVT), a relatively uncommon vascular complication of SLE.

CASE

A 17 year-old female was admitted to our department with flank pain, flank tenderness, fever, and arthralgia for two days. The patient had not been receiving any medications and there was no previous history of deep vein thrombosis or systemic disease. On physical examination, her blood pressure was 120/80 mmHg, pulse rate 82/min, and body temperature 37.8 ^{o}C. An oral aphthous ulcer and malar rash were detected. The history revealed photosensitivity. All examinations of other systems proved normal. Laboratory evaluation revealed serum urea 20 mg/dl (10-50), creatinine 0.98 mg/dL (0.6-1.3), total serum protein of 66 g/L (64-83), serum albumin 35 g/L (35-54), erythrocyte sedimentation rate 108 mm/h, hemoglobin 9.6 g/dl, white blood cell count 10.9 K/uL, and platelets 283.000/mm^3. Other hematological and biochemical parameters were normal. Automatic full urine test results were normal and urine microscopy revealed 6-7 erythrocytes per-high power field without any casts. 24-hour protein excretion was 400 mg. HbsAg, HBeAg, anti-Hbs, anti-Hbe, anti-HCV and HIV were all negative. Her

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Correspondence Address:
Ezgi ÇOŞKUN YENİGÜN
Numune Eğitim ve Araştırma Hastanesi,
Nefroloji Kliniği, Ankara, Turkey
Phone : + 90 312 508 45 52
E-mail : drezgi_76@hotmail.com

antinuclear antidody (ANA) test was positive but anti-double-stranded DNA (anti-ds-DNA) was weakly positive and her C3 level was normal. Anticardiolipin antibody, antiphospholipid antibodies, factor V Leiden, prothrombin gene mutation, activated protein C resistance were all negative and protein C and S were normal. Renal Doppler sonography revealed normal kidney size (right kidney:104 mm, left kidney 94 mm) and large thrombus formation at right renal vein with extension to inferior vena cava. Computed tomography and angiography revealed severe occlusion of the right renal vein without collateral vessels. She had 4 of 11 American Rheumatism Association (ARA) criteria for SLE, so she was diagnosed as SLE. RVT was treated with systemic thrombolytic therapy (streptokinase 1.5 million units intravenously over 60 minutes via peripheral intravenous catheter without any complication) for three successive days, and then with prednisolone (1 mg/kg/d), hydroxychloroquine and long-term (for six months) warfarin treatment with a target INR of 2.5–3.0. Control Doppler sonography revealed thrombus formation at the right renal vein while renal perfusion scintigraphy showed no perfusion or function of right kidney. The patient was diagnosed to have RVT associated with SLE.

DISCUSSION

RVT has been mostly shown in cases with nephrotic syndrome; but trauma, infection and malignancies are main causes of this vascular complication. Acute onset of RVT may also occur in association with anti-phospholipid syndrome (APS), injury to the renal vein during renal venography, trauma, postrenal transplantation, surgery around the renal vein, and occasionally in nephrotic syndrome (2). The incidance of RVT due to membranous nephropathy was shown 5-62% in literatures (3,4). Presence of anti-phospholipid antibodies (APA) is the most common cause of spontaneous RVT and they are found 35-40% of SLE patients (2)

SLE itself is associated with an increased incidence of venous thromboses and thrombophlebitis but venous thrombosis is slightly less frequent than arterial thrombosis in SLE. There is less information that the exact incidence of venous thromboembolism (VTE) in SLE, but a few studies showed that the incidence of VTE in SLE patients 5-10% patients (5-7). Beside the antiphospholipid antibodies, the antiprothrombin, anti-annexin V, anti factor XII and anti-protein S antibodies, factor V leiden, prothrombin gene mutation and acquired activated protein C resistance are reported other causative factors for VTE in SLE (7-9). In the study revealed that VTE in SLE occurred early , 50% of the events developing in the first 2.5 years, during the course of the disease. In younger patients especially less than 50 years of age, incidence of VTE in SLE was significantly higher (9).

The first RVT in a lupus patient has been reported in in 1968 (10). RVT in SLE is rare and the exact incidence is unknown. In the study has shown that the incidence of RVT was almost 2% in the kidneys of 100 patients with SLE (11).

The pathogenesis of RVT in SLE is not completely understood. Multiple factors might be indicated, including nephrotic syndrome (12), membranous glomerulonephritis (13), hypercoagulation state (14), prior episodes of thrombophlebitis (15) and anticardiolipin antibody (ACA) (16). As in other renal diseases, RVT in SLE appears to be a complication rather than a cause of the nephrotic syndrome. Gilsanz et al searched RVT by angiography in 20 SLE patients (17). RVT was detected in 2 of 6 patients with the nephrotic syndrome but only in 1 of 14 without nephrotic syndrome. However, nephrotic syndrome has been reported to be a distinct risk factor for RVT in SLE patients (18), a study showed that patients with peripheral thrombophlebitis had a high risk of developing RVT (61.5%) than with nephrotic syndrome (27%) (15). In contrast, our patient had no nephrotic syndrome, thrombophlebitis history or anticardiolipin antibody. The reason for this disparity is unknown but an ethnic difference is possible.

Imaging remains the cornerstone of diagnosis. Radiological signs of the affected kidney is an enlarged and hyper-echogenic kidney in approximately 90% of the patients in the early phase of acute RVT (19). To visualize the renal veins by CT anjiography can be used because non-invasive and high diagnostic accuracy. Simultaneous intravenous administration of contrast (CT angiography) assists in the visualization of the renal veins. The sensitivity and specificity of CT angiography is almost 100%.

The management of patients with SLE and RVT has been similar to that of RVT in other conditions. Although RVT in SLE is rare clinical condition, clinicians should be aware of this disease, especially in young women.

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