

IgA Nephropathy in a Patient with Systemic Lupus Erythematosus

Sistemik Lupus Eritematozlu Bir Olguda IgA Nefropatisi

ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune disorder. One of the most typical aspects of SLE is renal involvement. Lupus nephritis is a serious disease whose prognosis can usually be improved dramatically by treatment. The occurrence of non-lupus nephritis in SLE patients has rarely been reported. IgA Nephropathy (IgAN) and lupus nephritis (LN) have quite different laboratory, histopathologic findings and extra-renal clinical manifestations. In the light of these data we thought that IgAN and LN have different pathogeneses. We report a patient with SLE in whom IgA nephropathy was diagnosed.

KEY WORDS: IgA nephropathy, Lupus nephritis, Systemic lupus erythematosus

ÖZ

Sistemik lupus eritematoz (SLE) otoimmün bir hastalıktır. SLE'nin en tipik özelliklerinden birisi böbrek tutulumudur. Lupus nefriti ciddi bir hastalıktır ki prognozu genellikle tedavi ile önemli ölçüde geliştirilebilir. SLE hastalarında lupus nefriti dışı nefrit oluşumu nadiren bildirilmiştir. IgA Nefropatisi (IgAN) ve lupus nefritinin (LN) oldukça farklı laboratuvar, histopatolojik bulguları ve ekstrarenal klinik belirtileri vardır. Bu veriler ışığında IgAN ve LN'nin patogenezinin farklı olduğunu düşünmekteyiz. Burada biz IgAN'si tespit edilen SLE'li bir olgu sunacağız.

ANAHTAR SÖZCÜKLER: IgA nefropatisi, Lupus nefriti, Sistemik lupus eritematoz

INTRODUCTION

Autoimmune disorders might develop under the effect of various genetic, immunological, hormonal or environmental factors and they can involve a single organ or multiple organs and tissues (1). Systemic lupus erythematosus is a multisystem autoimmune disease with kidney involvement. In 50% of the patients, renal involvement can be clinically detected but this rate is higher in renal biopsy series (2). Clinically, Lupus Nephropathy (LN) has six types, which are minimal mesangial LN (class I), mesangial proliferative LN (class II), focal proliferative LN (class III), diffuse proliferative LN (class IV), membranous LN (class V), and advanced sclerosing LN (class VI) (3). The typical LN is characterized by so-called "full-house" pattern under immunofluorescent

microscopy when positively stained for IgG, IgA, IgM, C3, and C1q (4). Other than the typical subgroups, a few SLE patients were reported to have IgA Nephropathy (IgAN) established by renal biopsy (5).

The most commonly observed glomerulopathy is IgAN in developed countries (6). Glomerular capillary damage and diffuse IgA deposits in glomerular mesangium are morphological characteristics of the disease. The frequency of IgA deposition in glomerular mesangium is 5-15% in the overall population; despite this high ratio, these deposits result in clinical findings (such as hematuria/proteinuria) in only one out of every 50 persons (7). IgA nephropathy is an uncommon cause of proteinuria in lupus nephritis. In the case presented here, IgA nephropathy was observed in the renal biopsy of a patient with SLE.

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CASE REPORT

A 22-year-old female patient with complaints of pain in the joints of the knees and hands at the internal medicine outpatients was referred to our clinic to determine the cause of proteinuria. The patient's laboratory results were as follows: serum urea 14 mg/dl, creatinine: 0.7 mg / dL, total protein: 5.6 g/dl, albumin: 1.5 g/dl, triglycerides: 284 mg/dl, LDL: 174 mg/dL, HDL: 44 mg/dl, hemoglobin: 12 g/dl, Hct: 36%, WBC 8000/mm³, platelet 393000/mm³ sedimentation: 83 mm/h, CRP: 0.3, ANA:(+), antids-DNA(+3), IgG: 2440 mg/dl, IgA: 392 mg/dl, IgM: 261 mg/dl, rheumatoid factor: (-), C3: 79.5 mg/dl (79-152), C4: 10.8 mg/dl (16-38), HBsAg: (-), HCVab: (-), HIV:(-), TIT density: 1015, protein:(+3), microscopy: 1-2 leukocytes, proteinuria: 8648 mg/L/day.

The patient was consulted to the rheumatologists because of the malar rash, joint pains and laboratory results and was diagnosed as SLE. Kidney size and parenchymal thickness were normal on ultrasonographic measurement and renal biopsy was performed due nephrotic proteinuria. The biopsy specimen included 31 glomeruli and two of them globally had sclerotic changes but the others had no specific morphological feature. The interstitium and interstitial vessels were almost normal. Mesangial IgA deposits were positive under immunofluorescent microscopy investigation and trace amounts of anti C3 ab were also positive but anti C4 ab and anti C1q ab were negative. It was speculated that these findings are more common in IgA nephropathy than SLE nephritis.

Prednisolone 55 mg/day was prescribed and subsequently the dose was gradually reduced and cyclosporin 200 mg/day was added to the treatment. After the improvement of the patient's clinical and biochemical findings, the prednisolone and cyclosporine therapy was discontinued in the second year. The patient has normal blood pressure and no proteinuria on routine follow-up and is using hydroxychloroquine for the joint pains.

DISCUSSION

SLE is a systemic, autoimmune disorder that affects women, especially in their 20s and 30s. The most serious clinic form of SLE is lupus nephritis that it usually starts in the first 5 years of the disease. At the time of the diagnosis of lupus nephritis, an abnormal urinalysis with or without an elevated plasma creatinine concentration is present in a large proportion of patients and may develop in up to 50 percent of patients in time (2). Proteinuria is the most frequent abnormality observed in patients with lupus nephritis. Renal biopsy is an important tool to assess the degree of renal involvement as well its activity and damage, and can guide the therapeutic decision (8).

Typical extra-renal signs in the skin, joints and blood are often associated with LN, which allows easy differentiation from IgAN. On the other hand IgAN might have also some extra-renal signs, such as arthralgias, vasculitic type skin lesions

and erythema nodosum (9). The mesangial proliferation and mesangial deposits of IgA (usually absent in lupus nephritis), C3 fraction of complement and occasionally IgG and IgM, which are responsible for complement activation with the consequent release of inflammatory mediators, are characteristic immune histological features of IgAN. The absence of C4 and C1q deposits in LN (10) suggest that the alternative way of complement activation is involved in the pathogenesis of the disease.

Nevertheless, the laboratory and histopathologic findings of IgAN and LN and their extra-renal clinical manifestations are quite different and support a different pathogenesis. Typical LN is characterized by a "full-house" stain under immunofluorescent microscopy, staining positively for IgG, IgA, IgM, C3, and C1q. Only IgA staining in LN is a rare presentation. The occurrence of IgAN during SLE is also a rare event and, given the relatively high frequency of IgAN, could be attributable to a casual association, but considering the physiopathological mechanisms of both diseases it assumes pathological and therapeutic interest.

There is limited data on long-term treatment of IgA nephropathy. Cyclosporine has been investigated in small series of patients with IgA nephropathy (11). Our case also showed that CycA might be a therapeutic option for the treatment of the disorder.

In conclusion, SLE should be kept in mind in young women patients and the extrarenal manifestations of the disease must be questioned properly even though the patient was diagnosed as IgA nephritis with kidney biopsy.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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