İst Tıp Fak Derg 2009;72:105-107
J Ist Faculty Med 2009;72:105-107
www.itfdergisi.com

ERİŞKİN SİSTEMİK LUPUS ERİTEMATOZUS VE 2. DERECE ATRİYOVENTRİKÜLER BLOK

ADULT SYSTEMIC LUPUS ERYTHEMATOSUS AND SECOND-DEGREE ATRIOVENRICULAR BLOCK

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ÖZET

Dal blokları, birinci, ikinci derece ve tam kalp bloğunu içeren atriyoventriküler (AV) iletim bozukluğu, erişkin sistemik lupus eritematozusta (SLE) nadiren görülür. Burada standart doz steroid ile düzelen SLE erken bulgusu olarak değerlendirdiğimiz, Anti RNP, Anti-Ro antikorları pozitif 2. derece AV bloklu hastayı sunuyoruz.

Ellidokuz yaşında bayan yaklaşık 1 aydır olan yorgunluk yakınması ile başvurdu. Soy ve özgeçmişinde kardiyak hastalık dahil özellik tariflemiyordu. Sigara içmiyordu. Muayenesinde kan basıncı 120/80 mmHg, kardiyak nabız 62/dk ritmik, radiyal nabzı 62/dk ritmik idi. Malar rash, oral aftları vardı. Kalp oskültasyonunda üfürüm duyulmadı. Elektrokardiyografisinde (EKG) 2. derece atriyoventriküler Mobitz tip 1 blok tespit edildi. Antinükleer antikor 1/1000 granüler paternde, anti Sm, Ro antikorları pozitif tespit edildi. Anti-RNP antikorları 1/600 oranında pozitifti. Oral aft, malar rash, proteinüri, ANA pozitifliği ile SLE tanısı kondu. Metil prednison 1mg/kg başlandı. Tedavinin 5 gününde yorgunluğu azalan hastanın EKG'si normaldi.

Özellikle neonatalde iyi bilindiği gibi anti-Ro, anti-RNP antikorları pozitif tespit edilen erişkin hastalar kalp blokları açısından takip edilmelidir.

Anahtar kelimeler: Sistemik lupus eritematozus, ikinci derece atriyoventriküler blok

ABSTRACT

Systemic lupus erythematosus (SLE) rarely causes atrioventricular (AV) conduction disorders in adults including first- and second-degree bundle branch blocks and complete heart block. This report will present a case of second-degree AV block with anti-RNP and anti-Ro antibodies, an early sign of SLE, which improved with a standard dose of steroids. A fifty-nine-year-old woman presented with tiredness lasting for about one month. History did not reveal any cardiac illness. She was a non-smoker. On physical examination, she had a blood pressure of 120/80 mmHg, a rhythmic cardiac pulse of 62/min and a rhythmic radial pulse of 62/min. She had malar rash and oral aphthae. Cardiac oscultation did not reveal a cardiac murmur. Electrocardiography (ECG) showed Mobitz type I block, a second degree AV block. Antinuclear antibodies (1/1000) were in granular shape and anti-Sm and anti-Ro antibodies were positive. Anti-RNP antibodies (1/600) were also positive. The diagnosis of SLE was based on oral aphthae, malar rash, proteinuria and ANA positivity. Methyl prednison 1mg/kg was instituted. The patient noted that her fatigue subsided on the fifth day of the treatment and her ECG was normal.

Although anti-Ro and anti-RNP antibodies are known to be positive in neonates with AV block, adults with anti-Ro and anti-RNP positivity should also be followed carefully for heart blocks.

Key words: Systemic lupus erythematosus, second-degree atrioventricular block

INTRODUCTION

Systemic lupus erythematosus (SLE), an autoimmune disease, affects immune complexes and various tissues and organs through autoantibodies (8). It involves the heart in over 50% of the cases and the most frequently affected is the pericardium (16). SLE rarely causes trioventricular (AV) conduction disorders in adults including first- and second-degree, bundle branch blocks and complete heart block (1,2,6,7,9,11,14).

Date received/Dergiye geldiği tarih:16.12.2007 - Dergiye kabul edildiği tarih: 20.03.2008

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This report presents a case of second-degree AV block with anti-RNP and anti-Ro antibodies, an early sign of SLE, which improved with a standard dose of steroids.

CASE

A fifty-nine-year-old woman presented with fatigue lasting for about one month. History did not reveal any cardiac illness. She was a non-smoker. On physical examination, she had a blood pressure of 120/80 mmHg, a rhythmic cardiac pulse of 62/min and a rhythmic radial pulse of 62/min. She had malar rash and oral aphthae. Cardiac oscultation did not reveal a cardiac murmur. Laboratory investigations revealed that hemoglobin was 8.4 gr/dl, haematocritis 25,4% white blood cell count 2600, platelet count 79000, erythrocyte-sedimentation rate 79 mm/h, urea 80 mg/dl (normal range: 13 - 43), creatinine 1.3 mg/dl (normal range:0.6-1.1), albumin 2.7 g/dl (normal range:3.5-5), globulin 3.4 (normal range:2.9-3.3), and transaminases, lactic dyhdrogenase, calcium, sodium, potassium and magnesium values were normal. Electrocardiography (ECG) showed Mobitz type I block, a second degree AV block. Heart rate was about 60/min (Figure A, pretreatment). Creatinine, phosphokinase-MB and troponin I values were

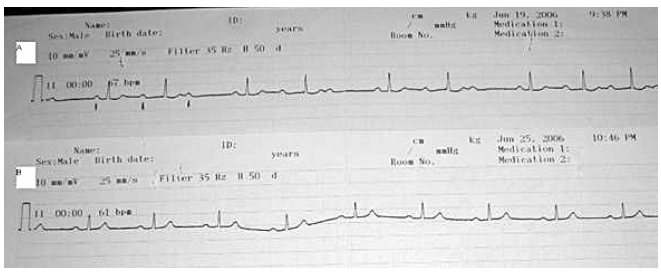

Figure A-B(Pre-post treatment): 2:1 second-degree atrioventricular block and normal ECG.

normal and the patient did not describe chest pain. Examination of the urine collected for 24 hours revealed proteinuria of 600 mg/day. Antinuclear antibodies (1/1000) were in granular pattern and anti-Sm and anti-Ro antibodies were positive. Anti-RNP antibodies (1/600) were also positive. Activated partial thromboplastin time was 28 seconds. Lupus anticoagulant test was negative. Complement (C3) was 79 mg/dl (normal range: 90-180 mg/dl). Anticardiolipin antibodies, Ig and IgM were negative. Echocardiography revealed minimal insufficiency of the aorta, and tricuspid and mitral valves. Ejection/fraction rate was 70%. Iron, serum iron binding capacity, iron saturation and vitamin B12 levels were normal, but ferritin was 1309 ng/ml (normal range:13-150). On peripheric blood film, neutrophils were 50%, lymphocytes were 44%, monocytes were 6%, erythrocytes were hypochromic, and there was anisothrombocytosis and clusters of 5-6 platelets. Reticulocytes were 1%. Evaluation of the bone marrow revealed minimal dysplasia in erythroid series. The diagnosis of SLE was based on oral aphthae, malar rash, proteinuria and ANA positivity (18). Methyl prednison 1mg/kg was instituted. The patient noted that her tiredness subsided on the fifth day of the treatment and her ECG was normal (Figure B, posttreatment). Ocular examination showed no abnormality. The patient was also given chloroquine 500 mg/day. One month later, her malar rash decreased, oral aphthae disappeared, complete blood count, urea and creatinine levels and ECG were normal. Erythrocyte-sedimentation rate was 44/mm/h.

DISCUSSION

Clinical signs, laboratory investigations or autopsy may reveal involvement of the pericardium, myocardium, cardiac valves, conduction system and coronary arteries caused by SLE. Anticardiolipin antibodies may account for cardiac involvement (6,7,16).

Godeau et al. (6) in their series of 103 SLE patients reported that 14.5% of SLE cases had conduction disorders. AV blocks have been reported to appear late, present with acute exacerbations of the disease and can be associated with anti-malarial drugs. Godeau et al. (7) in their series of 112 SLE patients found 18 patients (17.5%) to have conduction disorders. Of 18 patients, five had right bundle branch block, 2 complete heart block, three partial left bundle branch block, two first-degree AV block and one second-degree AV block. They followed the cases of AV block for 9-20 years and reported that conduction defects appeared more frequently than they expected, developed during the course of the disease and could be caused by malarial drugs. We also observed AV block as the first sign of SLE and the patient had not received any treatment yet.

James et al. (10) attributed conduction defects to the presence of pericarditis, arteriopathy, involvement of the endocardium below the sinus node and collagen abnormality of the AV node. The case presented here did not have pericarditis. Since electrocardiography and cardiac enzymes did not suggest a coronary artery disease, coronary imaging was not performed.

AV blocks are rare in adults with SLE, though they frequently appear in the neonates with SLE. AV blocks in the neonates are known to be associated with anti-Ro antibodies (3). The case presented in this report also had anti-RNP and anti-Ro antibodies. It is not clear whether anti-RNP and anti-Ro antibodies lead to AV block in adults with SLE. Logar et al. (13) in their prospective study on 67 SLE patients reported that one patient had first-degree AV block and three patients had first-degree AV block accompanied by unifascicular block and SSA/Ro antibodies. They did not observe second-degree AV block. AV conduction defects can be associated with lupus exacerbations. O' Neill et al. (17) in their series of 43 SLE patients claimed that conduction defects were not associated with anti-Ro antibodies. However, there have been a lot of case reports which put emphasis on an association of conduction defects with anti-RNP and anti-Ro antibodies (1,2,9,11,14). In recent years, Anti-Ro antibodies have been shown to interrupt AV conduction in fetal cardiocytes through an influx of calcium (15). Ro antigens have been shown to be present in nuclei of the myocardial cells and it has been associated with cardiac block (4) .

Second-degree AV block in a case of mixed connective-tissue disorder has been associated with anti-RNP antibodies (19). However, the case presented here did not have mixed connective-tissue disorder because she had anti-RNP titers of below 1/1000 and did not have the clinical signs of the disease such as edema of the hands, Raynaud's phenomenon, acrosclerosis, synovitis, and myositis.

It has been reported that anti-Ro is positive in cases of high degree blocks including second-degree AV block (2,11,14). Some authors have emphasized the presence of anti-RNP antibodies (1,2,5,9,11). One case of SLE has been reported to have second-degree AV block associated with anti-RNP and anti-Ro antibodies (11). The case has been observed to develop first-degree AV block at first and then second-degree AV block. First-degree AV block has been reported to subside with a high dose of steroids and ECG has been normal on the third day of treatment. A second-degree AV block was observed in this case and the patient had normal cardiac rhythm on ECG on day five.

Some authors have underlined that a bundle branch block, already present, may cause cardiac block (1,2). Godeau et al. reported cases of heart block associated with or without a bundle branch block (6,7).

Although there has not been a specific treatment approach for AV block, high doses of steroids have been used for the treatment of high degree AV blocks (6,19). 1mg/kg steroids were initiated and the patient responded to the treatment. High degree blocks may require treatment with a pacemaker (2,7).

Although anti-Ro and anti-RNP antibodies are known to be positive in neonates with AV block, adults with anti-Ro and anti-RNP positivity should also be followed carefully for heart blocks.

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