Acute rheumatic carditis associated with Schoenlein-Henoch vasculitis

Schoenlein-Henoch vaskülitiyle birlikte gelişen akut romatizmal kardit

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Introduction

In 1948 Gairdner firstly defined the linkage of Schoenlein-Henoch purpura (HSP) vasculitis with rheumatic fever (1). Since the first report of that association, only a few cases of HSP with acute rheumatic fever have been reported (2-5). In this article, we present a patient admitted to our clinic with clinical picture of HSP vasculitis, who developed acute rheumatic carditis thereafter.

Case Report

An 8 years old girl was admitted to our clinic with the complaints of fever and a 3 days history of purpuric rash on her lower extremities. Her history revealed that she had an upper respiratory infection 10 days ago. At the time of admission her axillary body temperature was 37.7°C. She had exudative tonsillo-pharyngitis, widespread petechial and purpuric rash on her lower extremities. Her cardiac auscultation was normal. Her initial laboratory findings showed no pathology except slightly elevated erythrocyte sedimentation rate, 48 mm/hour and leucocyte count, 13.200/mm^3. Penicillin intramuscularly was given and her control examination performed on the third day of penicillin therapy revealed she had still high fever 38.5°C, the rash had been spread over the buttocks and abdominal pain developed. She had diffuse arthralgia and her both ankles were swelled. The most striking finding was a systolic cardiac murmur of 2/6 grade best heard in the apical region. Her electrocardiography and telecardiography were normal. Echocardiographic findings revealed moderate to severe mitral valve regurgitation. Her C-reactive protein was elevated, 13.5 mg/dl and antistreptolysin (ASO) titer was 631 Todd units. Leucocyte count was 32.000/mm^3, erythrocyte sedimentation rate was 54 mm/hour. Urine analysis showed no proteinuria or hematuria, and urine culture was negative for bacteria. Stool examination was negative for occult blood. Serum immunoglobulin (Ig) A level was elevated (296 mg/dl, normal range: 33-236 mg/dl) that was consistent with HSP vasculitis. Anti-nuclear antibody was absent, rheumatoid factor was negative, complement 3 (83 mg/dl, normal: 77-195) and complement 4 (25 mg/dl, normal: 7-40) levels were normal. The skin biopsy showed leucocytoclastic vasculitis. Her throat culture was negative for group-A beta-hemolytic streptococcus (GABHS) infection. By those findings, HSP vasculitis with acute rheumatic carditis was diagnosed and prednisone 2 mg/kg/day p.o. was started.

Discussion

HSP vasculitis and acute rheumatic fever are both multisystemic diseases affecting joints and heart. Our patient had migratory arthritis and valvulitis, which favored acute rheumatic fever. Arthritis and arthralgia may be found in HSP, but arthritis in HSP patients is non-migratory. It is proposed that antigenic stimuli of group-A beta-hemolytic streptococcus (GABHS) infections may trigger HSP (6). In previous reports association of HSP vasculitis with GABHS infection had been emphasized, in our case throat culture was negative for group A beta-hemolytic streptococcus because of the former antibiotic usage, since it was taken after penicillin therapy. However, elevated ASO titers suggested a recent streptococcus infection.

Different immunological mechanisms are responsible for the pathogenesis of HSP vasculitis and acute rheumatic fever. Cellular immune response mediated by T-lymphocytes to streptococcal M-proteins cause destruction of cardiac tissues (7). HSP vasculitis is an IgA mediated disorder, raised serum IgA levels, IgA class antibodies and immune complexes have been described in those patients (8). In our patient serum IgA level had been elevated and skin biopsy demonstrated leucocytoclastic vasculitis which was consistent with HSP vasculitis. Even though the role of GABHS infection in HSP vasculitis isn't clear, GABHS antigens may initiate parallel immunological processes resulting in both disorders.

Cardiac involvement in HSP vasculitis patients is very rare and previously there was only one report of HSP and acute rheumatic fever co-occurrence without time interval between those disorders (9). In a previous report, the interval from onset of rash to appearance of rheumatic fever was declared as 10 days to 12 weeks (10). Our patient was firstly diagnosed as HSP vasculitis and 3 days after, a cardiac murmur of acute valvulitis and migratory arthritis developed. She was the second case who developed acute rheumatic fever at the same time with HSP vasculitis in current literature.

Conclusion

Since there were a few reports concerning uncommon co-occurrence of HSP and acute rheumatic fever, we aimed to report that case. Our patient was the second presented case with acute rheumatic fever associated with HSP vasculitis with no time interval.

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