

Systemic Lupus Erythematosus and Hyper-eosinophilic Syndrome An Unusual Association

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ABSTRACT

A 34-year old woman with dizziness, headache and both upper and lower extremities weakness was admitted to hospital. She had a history of photosensitivity but no asthma or allergy. On physical examination, malar rash and livedo reticularis were noted. White blood cell count was 18500/ μ L with 7585 eosinophils (41%). She also had positive antinuclear antibody (ANA), anti-double stranded DNA antibody (anti-ds-DNA antibody) and anticardiolipin antibody (aCL antibody). Echocardiography revealed left and right ventricular obliteration with fibromatous biventricular endothelial thickening. Brain MRI showed changes in favour of white matter ischaemia and lacunar infarction. Hyper-eosinophilic syndrome (HES) and systemic lupus erythematosus may be considered to have occurred concurrently in this patient.

El Lupus Eritematoso Sistémico y el Síndrome Hipereosinofílico Una Asociación poco Usual

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RESUMEN

Una mujer de 34 años de edad con mareos, dolor de cabeza y debilidad en las extremidades superiores e inferiores, fue ingresada a nuestro hospital. La mujer tenía una historia de fotosensibilidad pero no de asma ni alergia. En el examen físico, se observó erupción malar y livedo reticularis. El conteo de glóbulos blancos fue 18500/ μ L con 7585 eosinófilos (41%). También tenía anticuerpos antinucleares (ANA) positivos, anticuerpos anti-DNA de hebra doble (Anticuerpos anti-dsADN), y anticuerpos anticardiolipina (anticuerpos aCL). La ecocardiografía reveló obliteración ventricular izquierda y derecha con engrosamiento endotelial fibromatoso biventricular. La resonancia magnética (MRI) del cerebro mostró cambios a favor de la isquemia de la materia blanca y el infarto lacunar. Puede considerarse que el síndrome hipereosinofílico (SHE) y el lupus eritematoso sistémico tuvieron lugar de modo concurrente en este paciente.

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INTRODUCTION

The hyper-eosinophilic syndrome (HES) is marked by a sustained overproduction of eosinophils (1). Currently present evidence show that HES presents with symptoms such as fatigue, cough, breathlessness, muscle pain, angioedema, rash, fever and retinal lesions (2). Herein, we describe a young lady with non-erosive arthritis who later developed eosinophilia with end organ involvement. She also had positive antinuclear antibody (ANA), anti-double stranded DNA antibody (anti-ds-DNA antibody) and anticardiolipin

antibody (aCL antibody) titres which to the best of our knowledge have rarely been reported together.

Case Report

The patient was a 34-year old woman referred to the Rheumatology Ward of Hafez hospital, Shiraz University of Medical Sciences with drowsiness, dizziness, headache and generalized weakness. She had been followed-up by a private rheumatologist for some years for arthritis in both upper and lower extremities, and had been prescribed prednisolone and methotrexate. She also had a history of hair loss and photosensitivity, but no asthma, pruritic skin rashes, rhinorrhoea or any other complaint attributable to allergic disorders, and was on no medication for the last 24 months.

On admission, she had malar rash without any abnormality in the lung fields, heart and abdomen. Periungual

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erythema and *livedo reticularis* were detected in both feet. Motor powers were grade 4/5 in both upper and lower extremities. The white blood cell counts were 18500/ μ L comprising 7585/ μ L (41%) neutrophils, 2590/ μ L (14%) lymphocytes, 740/ μ L (4%) monocyte and 7585/ μ L (41%) eosinophils.

During four weeks of hospitalization, the following tests or procedures were performed. She had negative serology for *Toxocara catis* and *canis*, normal creatinine phosphokinase and lactic dehydrogenase, a positive ANA with a homogenous pattern, a positive anti-ds-DNA antibody, a positive aCL antibody of 55 GPL unit/ml and a low C4 component of complement. Antineutrophilic cytoplasmic antibodies (C-ANCA and P-ANCA) were negative and serum IgE level was in the upper normal limit. Stool examination by concentrated method showed no parasite. The synovial fluid examination showed no cell with normal protein and glucose and the result of the bone marrow aspiration and biopsy was normocellular marrow without any pathology. Nerve conduction velocity and electromyography tests were normal. Echocardiography showed left and right ventricular apical obliteration with fibrothrombus, biventricular endocardial thickening. Restrictive left ventricular filling, mild mitral valve regurgitation and mild pulmonary hypertension were also noted (Fig. 1). Brain MRI revealed multiple hy-



Fig. 1: The findings of echocardiography. Left and right ventricular apical obliteration, with fibrothrombus biventricular endocardial thickening. Restrictive left ventricular filling, mild mitral valve regurgitation and mild pulmonary hypertension.

perintense lesions in the white matter, some with central hypointensity suggesting white matter ischaemic changes and small lacunar infarctions in parietal lobes and at the level of the centrum semiovale (Fig. 2). Fluorescent angiography showed thrombosis in the right retinal artery.

The patient was treated with intravenous pulse methylprednisolone and therapeutic doses of heparin. Ten days later, the motor powers of both upper and right lower ex-

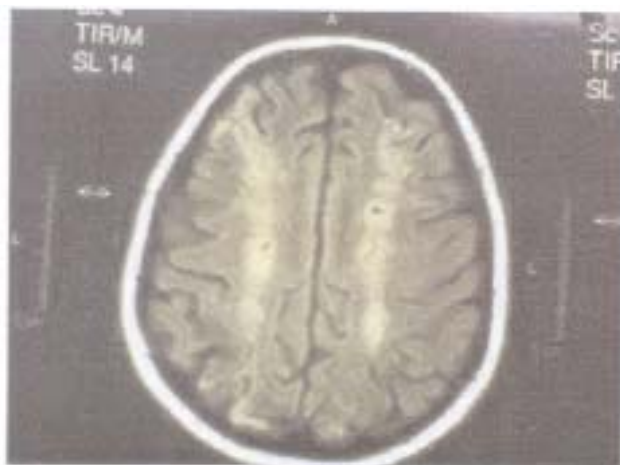


Fig. 2: The findings of brain MRI. Multiple hyperintense lesions in the white matter, some with central hypointensity suggesting white matter ischaemic changes and small lacunar infarctions in parietal lobes.

trémities improved to 5/5. The WBC and eosinophil counts were 8400/ μ L and 588 / μ L respectively. On follow-up in the outpatient clinic, she still had left lower extremity weakness (4/5). No malar rash, *livedo reticularis* or *periungal erythema* was found. Echocardiography showed mild mitral regurgitation and mild pulmonary hypertension. She was on 5 mg prednisolone every other day, warfarin (5 mg) and azathioprine (50 mg) per day as a steroid sparing agent.

DISCUSSION

Although any organ may be involved, the heart, skin, nervous system, lung and spleen have been reported to be involved in 40 to 60% of patients with HES (3). Cardiac manifestations of HES include endocardial fibrosis and cardiomegaly (4). Pericardial effusion occurs rarely with HES (5–6). The syndrome may also be associated with cerebral thromboemboli, encephalopathy or peripheral neuropathy (7). Cerebral thromboemboli might occur in conjunction with cardiac involvement (7).

The present patient had endocardial thickening. Considering the reported association of cerebral thromboemboli and cardiac involvement (7), it might be possible to suggest that multiple bilateral lesions in the brain were due to embolic phenomenon which originated from the endocardial thickening. Alternatively, they might be a result of an association with increased levels of ANA and aCL antibody.

Hyper-eosinophilic syndrome and SLE have been reported to occur concurrently (8). As far as the literature is concerned, this will be the first case report of simultaneous HES and SLE presenting with neurologic involvement, increased levels of aCL antibody and decreased levels of complement. Such a speculation is supported by the patient's symptoms including previous arthritis, malar rash, photosensitivity, central nervous system involvement, positive ANA, positive anti-ds-DNA antibody and aCL antibody, all of which point to SLE.

In conclusion, the clinical and laboratory findings indicate that HES and SLE had occurred concurrently in the present patient.

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