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ARTICLE

Report of Lupus Encephalopathy in Two Patients with Systemic Lupus Erythematosus

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ABSTRACT

How do you make a diagnosis of lupus encephalopathy (LE) in patients with systemic lupus erythematosus (SLE) concurrent with neurological and psychiatric abnormalities? This is a challenging question. We encountered two cases of SLE associated with LE. One involving the rigidity of the lefs due to extraparamidal vasculitis while the other involved mental illness due to frontal lobe atrophy respectively. The patient in case 2 experienced the attacks of Renault's sign, suggesting a systemic vasculitis. A final diagnosis of LE was established on the basis of elevated concentration of anti-ds-DNA in serum. The authors realized that the neurological or psychiatric symptoms in patients with rheumatic diseases should be carefully recognized in order to avoid a missed diagnosis of LE.

Keywords: Systemic lupus erythematosus (SLE); Lupus encephalopathy (LE); anti-ds-DNA; Dysfunction of brain

1. Introduction

We encountered two cases of systemic lupus erythematosus (SLE) that occurred in 81-year-old male and 71-year-old female respectively, who complained of rigidity of his legs for 16 years in case 1; and mental illness over the past two months. The patient in case 2 suffered a few attacks of "Renault's sign", suggesting a systemic vasculitis. Both of them showed a massive proteinuria in their urine samples. In their laboratory findings of immunology, a characteristic elevation in the concentration of anti-ds-DNA was demonstrated after admission in both cases. As an adjuvant, a single dose of "glucocorticoid" (GC) for diagnostic trial was also applied to help with the diagnosis for LE in case 2, and her psychiatric abnormalities mentioned below were alleviated steadily since then.

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2. Case Report

2.1 General Material

Case 1 The patient is a 81-year-old male, retired advanced accountant, who was admitted to the hospital with a chief complaint of difficult walking with an increased tension of muscle in both lower limbs over the past 16 years associated with a mild cognitive decline, drooling, kyphosis, right hip joint pain, and facial edema; which had been diagnosed as "Parkinson's disease" (PD) by the Department of Neurology in a provincial hospital, and has been taking "antiparkinsonian drugs", such as Madopar, Pramipexole. The effect for PD has declined in recent years. Due to his "diversity" of the symptoms (involving multiple organs), the physicians expanded the relevant tests. A positive anti-ds-DNA was found, followed by a heavy β2- microglobulinuria which occurred at 25 times higher than normal limits. Further X-ray examination of his lumbar spine and right hip joint showed the signs of "ankylosing spondylitis"; Six items of his cellular immunity were also abnormal, and the final diagnosis of SLE was established concurrent with LE, in which the "extrapyramidal system" was damaged, leading to secondary Parkinson syndrome (PS). As a result, integrated therapy of TCM and western medicine for immunosuppression was given for his treatment.

Case 2 The patient is a 71-year-old female, retired entrepreneur, who was admitted to our hospital complaining of refractory eczema for one month. On the night of admission, the patient suddenly got up in the mid of the night and wanted to go home to get some clothes stubbornly. We conducted a MRI and the brain imaging showed atrophic frontal lobe and signal abnormality of temporal lobe. The patient show signs of "Renault's symptoms" as her left foot turns from pale to purple and redness. Her pretibial and ankle edema were obvious on both sides, together with headache attacks and being sleepy at times, she was also regarded as having "weird" behavior. Her urinalysis for microproteinuria showed 4 times higher than normal limit after admission, suggesting an early sign of renal involvement. The ratio of CD4/CD8 was 2.8 (normal value: less than 2.5) in her immunological test, indicating her disturbed immunity. A "butterfly erythema" on her cheeks was also found during a ward round, and a single dose of "glucocorticoid" (GC) for diagnostic trial was subsequently given, relieving the abnormalities mentioned above. The diagnosis of SLE was finally confirmed concurrent with LE, which was the reason why the lady was presented with all those psychiatric symptoms.

3. Discussion

These were 2 cases of LE in our department of rheumatology in recent months, which were involved in the central nervous system (CNS), but they presented diversely. Case 1 showed elevated muscle tension on his extremities due to his "extraparamidal symptoms" (ataxia), together with "massive proteinuria" (facial and ankle edema). His diagnosis of PD lasting for 16 years had been made until the detection of anti-ds-DNA (+) this time, the main diagnosis was then corrected. Case 2 demonstrated "depression symptom" in her "frontal lobe syndrome," here means, a decreased restraint for motivation, and was suspected as "mental disorders". However, she was also accompanied by "massive albuminuria", attacks of "Reynolds's sign" and a positive antinuclear antibody (anti-ds-DNA); we established the diagnosis of SLE concurrent with LE on the basis of "a butterfly erythema" on her cheeks.

Lupus encephalopathy is defined as a specific type of SLE with CNS involvement, which can present with neurological and psychiatric disorders ; in other words, 8% -16% of SLE victims have vascular lesions (cerebral vasculitis) according to literature. There is a similar pathogenesis of vasculitis in brain to that of general vasculitis; i.e. Circulating Immune Complex (CIC) deposits and / or sensitized T-cell attacks Intracranially. The patient in case 2 show signs of episodic headache and "Renault's sign" in her left foot, this was evident to indicate that the patient might be suffering from systemic vasculitis (Pasquale et al., 2020)[1].

The process of two cases of "difficult diagnosis" in this paper suggested that the symptoms of rheu-

matic diseases are "systemic" in their features, that is, being characteristic of "multi-organ" damages (Lauren & William, 2020) [2]. Take case 1 as an example, when his rigidity of legs occurred for the first time, it is bound to register "neurological department" first, which would be a coincidence that the doctor who happens to treat the patient is good at PD, but not familiar with SLE. This "dilemma" needs to be solved by means of training more clinicians with the background of "integrative medicine" (Muller et al., 2008)[3]. The duty of rheumatologists is to probe into the "radical cause" behind the numerous symptoms, and also to the essence to carry out "immunosuppressive therapy" in a targeted manner to improve the efficacy.

4. Conclusions

We realized that a key to diagnosing LE in patients with SLE is about the relatively characteristic elevation of concentration of anti-ds-DNA in serum, integrated with neurological or psychiatric symptoms; which would be confirmed if we apply diagnostic trial of glucocorticoid to those suspected patients, as well as those classic measurements of immunology.

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