



Glucocorticoid reduction induced chorea in pediatric-onset systemic lupus erythematosus: A case report

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Abstract

BACKGROUND

Pediatric-onset systemic lupus erythematosus (SLE) is typically more severe than adult-onset SLE, with a higher incidence of nervous system involvement. Chorea is a relatively rare neurological complication reported in 2.4%-7% of SLE patients. In particular, chorea induced by glucocorticoid dose reduction is even rarer. Herein, we report the case of a girl with SLE, who developed chorea during the process of glucocorticoid therapy reduction.

CASE SUMMARY

We describe a 14-year-old girl who was diagnosed with SLE. She was treated with methylprednisolone and rituximab, and her symptoms improved. On the second day after the methylprednisolone dose was reduced according to the treatment guidelines, the patient developed chorea. Her condition improved after adjusting her glucocorticoid regimen.

CONCLUSION

This case is a reminder that extra attention to chorea is required in SLE patients during glucocorticoid dose reduction.

Key Words: Glucocorticoid; Chorea; Pediatric systemic lupus erythematosus; Case report

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Core Tip: Chorea can be induced by glucocorticoid dose reduction in patients with pediatric-onset systemic lupus erythematosus (SLE). Patients with SLE should be closely monitored during glucocorticoid reduction.

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INTRODUCTION

Pediatric-onset systemic lupus erythematosus (SLE) is generally more severe and has a higher incidence of neurological involvement than adult-onset SLE. The most prevalent neuropsychiatric syndrome manifestations include headache, cognitive dysfunction, and mood disorders[1]. Chorea is a relatively rare neurological complication reported in approximately 2.4%-7% of SLE patients (Table 1), and is rarer in SLE patients undergoing glucocorticoid therapy reduction. We report a pediatric patient with SLE, who improved after treatment with glucocorticoid and immunosuppressants. When the patient's condition was assessed as stable, the glucocorticoid was reduced according to routine treatment. Unfortunately, the glucocorticoid reduction induced chorea. With an increase in glucocorticoid dosage, the symptoms of chorea improved. This case is a reminder that extra attention to chorea is required in pediatric-onset SLE patients during drug dose reduction.

CASE PRESENTATION

Chief complaints

A 14-year-old female with fever, facial erythema, facial ulcers, mouth ulcers and alopecia was admitted to our rheumatology department.

History of present illness

Two months before admission, the patient began to develop facial ulcers with swelling, and no improvement was observed following treatment with clindamycin and metronidazole. Fever occurred one month before admission, and routine blood examination showed that the white blood cell count and red blood cell count were decreased. C-reactive protein (CRP) was normal, erythrocyte sedimentation rate (ESR) was 96 mm/h, and her body temperature continued to rise after dexamethasone and cefuroxime administration. One week previously, facial erythema accompanied by gaze (Video 1), fever, facial and oral ulcers, ulceration and bleeding on the ulcer surface, internal and external genital ulcers, severe hair loss, pharyngitis, and erythema on the fingertips of both hands and toes were observed.

Physical examination

A body temperature of 37.6 °C, blood pressure of 108/76 mmHg, heart rate of 126 beats/min, and respiratory rate of 20 times/min were noted. Ulceration and scabbing can be seen in the oral cavity. Facial erythema, ulcers at the fingertips of both hands and toes, vulvar ulcers and scar alopecia were observed. No joint tenderness. The remaining physical examinations showed no significant abnormalities.

Laboratory examinations

The results of laboratory examinations were as follows: ANA1: 1000, natural anti-SSA antibody, anti-dsDNA antibody, anti-nucleosome antibody, anti-cardiolipin antibody, and P-ANCA were positive. CRP was 9.78 mg/L, ESR was 68 mm/h, complement 3 (C3) was 0.11 g/L, and complement 4 (C4) was 0.01 g/L. The white blood cell count, red blood cell count, platelet count and hemoglobin were decreased, and a bone marrow biopsy showed no abnormalities. Respiratory virus, Mycoplasma infection, and procalcitonin results were negative. Given the patient's clinical symptoms, she was diagnosed with SLE. The Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2000) was 13 and we evaluated the activity as medium level. Following intravenous methylprednisolone 80 mg/d administration over five days and rituximab 500 mg once, the child's symptoms improved. C3 and C4 increased (C3 0.29 g/L, C4 0.02 g/L) and routine blood levels were normal. CRP and ESR decreased to within the normal range. The SLEDAI-2000 was 2. On the sixth day, methylprednisolone was reduced to 60 mg/d. The patient's body weight was 60 kg. The dosage of glucocorticoid as prednisone was 1.25 mg/kg/d due to decreased disease activity. Unfortunately, new symptoms emerged after one day of glucocorticoid reduction, her limbs and head exhibited involuntary dance-like movements, such as, turning of the neck, intermittent flexion and extension of fingers, waving the hand, and stretching of the arm (Videos 2 and 3). Further examination of the cerebrospinal fluid (CSF) including CSF biochemistry, next-generation sequencing of CSF, and autoimmune encephalitis antibody were negative. Anti-streptolysin O was negative. Thus, infectious encephalitis and autoimmune encephalitis were excluded.

Imaging examinations

Cranial magnetic resonance imaging revealed acute cerebral infarction in the left centrum semiovale.

Table 1 Prevalence of chorea in systemic lupus erythematosus

Year	Study	Prevalence
1987	Chorea in systemic lupus erythematosus and "lupus-like" disease: association with antiphospholipid antibodies	2.4%
2001	The prevalence of neuropsychiatric syndromes in systemic lupus erythematosus	2%
2002	The incidence and prevalence of neuropsychiatric syndromes in pediatric onset systemic lupus erythematosus	7%
2006	Neuropsychiatric manifestations in pediatric systemic lupus erythematosus: a 20-year study.	7%
2007	Neuropsychiatric involvement in pediatric systemic lupus erythematosus	Approximately 5%

FINAL DIAGNOSIS

Based on the patient's manifestations which included involuntary dance like movements of her limbs and head, and positive anti-cardiolipin antibody, she was diagnosed with chorea.

TREATMENT

As the SLEDAI-2000 changed to 27, we decided to administer more glucocorticoid. The patient received methylprednisolone (80 mg/d), intravenous immunoglobulin (20 g/d) for 3 d, cyclophosphamide (0.2 g every other day), low molecular weight heparin (2050 IU/d), sertraline (25 mg/d), and olanzapine (5 mg at night).

OUTCOME AND FOLLOW-UP

The patient's chorea symptoms improved significantly and she was able to move normally and answer questions clearly (Video 4).

DISCUSSION

SLE is an invasive autoimmune disease characterized by multiple system injury including the central nervous system. Among the neurological manifestations, chorea is rare in pediatric SLE patients[2]. The distinguishing feature of SLE is the production of autoantibodies, with the formation of immune complexes that precipitate at the vascular level, causing organ damage[3].

Previous studies have found that chorea can appear at any time in pediatric SLE, during the entire process of disease activity and clinical remission, and even several months after the onset of symptoms[4]. Glucocorticoid is the most important therapeutic agent in SLE patients with chorea. In this case, we treated pediatric lupus erythematosus with the standard glucocorticoid dosage. As the patient's condition improved, we gradually reduced the dosage of glucocorticoid according to the treatment guidelines[5]. However, the patient developed chorea symptoms on the second day of glucocorticoid reduction. As reported in the literature[6], the occurrence of chorea in SLE may be caused by the action of immune complexes, cytokines, and antiphospholipid antibodies (aPL) on cerebral blood vessels, which cause vascular occlusion, and corresponding brain dysfunction. Glucocorticoid dose reduction may lead to intensification of the inflammatory storm, and an increase in immune complexes which block cerebral blood vessels. Therefore, SLE should be fully re-evaluated when the glucocorticoid dose is reduced.

Chorea in SLE has been strongly associated with aPL. The positivity of aPL in SLE patients varies from 12% to 30%[7]. In a 50-patient study, among patients with positive aPL and chorea, 58% were diagnosed with SLE[8]. The pathophysiology of positive aPL with chorea in SLE has not been clearly identified. It may be involved in activation of aPL leading to neuronal disorders. aPL may pass through the blood-brain-barrier and connect to unknown antigens in the central nervous system, or to microthrombi that disturb local circulation in small cerebral vessels, causing disruption of the blood-brain-barrier, which normally protects the central nervous system from plasma proteins and cells[9]. Glucocorticoid dose reduction may lead to re-activation of aPL and result in brain thrombosis.

CONCLUSION

The emergence of chorea can be induced by glucocorticoid dose reduction in patients with pediatric-onset SLE. As shown by our case, we remind clinicians that when SLE patients have positive aPL, they should be particularly careful when reducing the dosage of glucocorticoid.

FOOTNOTES

Author contributions: Xu YQ contributed to the collection of medical record data and writing the first draft; Wang M and Zhang Y contributed to critically reviewed the paper; all authors actively contributed to the writing and reviewing of the article and approved the final version.

Informed consent statement: This study was conducted with the approval of the ethics committee of Chongqing Hospital of Traditional Chinese Medicine (approval No. 2023-GAKY-KS-XYQ). The patient gave explicit informed consent to report her clinical case.

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