Neuropsychiatric Manifestations in Pediatric Systemic Lupus Erythematosus

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Background: *The frequency of neuropsychiatric manifestations in systemic lupus erythematosus (NPSLE) is rare.*

Objective: *To determine the prevalence and clinical features of pediatric NPSLE.*

Material and Method: *We reviewed the medical records of pediatric SLE patients between January 1988 and August 2009. Sixty-six SLE patients were identified. The patients who had lupus-like symptoms and had NPSLE secondary to other causes were excluded.*

Results: *NPSLE was identified in 17 patients (25.8%). The median age onset was 13 years. Eight patients (12.1%) had NPSLE on initial diagnosis of SLE. Ten patients (58.8%) developed NP symptoms within the first year of SLE, while seven patients (41.2%) developed these symptoms after one year of SLE diagnosis. The most common symptoms were seizure (52.9%), stroke (29.4%), and movement disorder (17.6%). No laboratory finding was correlated with NPSLE. Brain imaging showed abnormalities in more than 50% of patients including infarction, vasculitis, brain atrophy, and subdural effusion.*

Conclusion: *The present study demonstrated that most of NPSLE patients developed symptoms within the first year after diagnosis. Interestingly, the prevalence of seizures in Asian pediatric SLE was found to be more significant than non-Asian population. We hypothesized that ethnicity may be one of the potential predictors of NP manifestations.*

Keywords: *NPSLE, Neuropsychiatric, Pediatric systemic lupus erythematosus*

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Systemic lupus erythematosus (SLE) is an autoimmune disease with multi-organ involvement. The neuropsychiatric (NP) manifestations in systemic lupus erythematosus (NPSLE) are quite variable and there were only a few studies focusing on children due to the lack of standard definitions. In 1999, the American College of Rheumatology (ACR) revised a standard nomenclature and a set of case definitions for 19 NP syndromes of NPSLE[14] in adult that also applied in children. The prevalence of NPSLE was reported to be 14% to 80% in adults[1-4,6,7,9,10], and varied from low 20% to high 95% in children[5,8,10,11,13,16,19]. In Thailand, there were only few studies on pediatric systemic lupus erythematosus[12,13] and no previous studies identified characteristic of NP manifestations. The aim of the present study was to determine the prevalence of NPSLE, clinical features, diagnostic evaluation, possible predictive factors, and treatment of NPSLE in pediatric SLE in Thailand.

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Material and Method

We retrospectively reviewed medical records of pediatric SLE patients diagnosed according to the ACR[32] that attended pediatric neurology clinic and nephrology clinic at Phramongkutklao Hospital between January 1988 and August 2009. We recruited all patients who were under 18 years of age at the time of onset of SLE. The study was approved by the Ethic Committee of Phramongkutklao Hospital. NPSLE were defined according to the Revised ACR nomenclature and case definitions for 19 NP syndromes of NPSLE[14]. We excluded the patients who had lupus-like symptoms from drugs and mixed connective tissue disease and had NPSLE secondary to other causes, such as hypertensive encephalopathy, uremia, infection, and other central nervous system (CNS) disease not related to SLE.

Demographic data, clinical features, diagnostic evaluation, and treatment of pediatric SLE patients were assessed. We reviewed laboratory findings at the time of SLE diagnosis including complete blood count, erythrocyte sedimentation rate (ESR), serum C3 and C4 levels, anti-nuclear antibody, anti-dsDNA antibody, anti-phospholipid antibody

(aPL), anti-cardiolipin antibody (aCL), pathological results of renal biopsies. Cerebrospinal fluid (CSF) analysis, computed tomography (CT), magnetic resonance image (MRI), and electroencephalography (EEG) results were performed at the time of diagnosis.

Statistical analysis

Data was analyzed using descriptive statistics and described in percentage, and median (range). Comparisons between patients with and without NPSLE were analyzed by using Mann-Whitney U test for continuous data and Chi-square or Fisher's exact test for categorical data as appropriate. The level of statistical significance for differences was less than 0.05.

Results

The clinical features of 66 pediatric SLE patients were summarized in Table 1. There were 52 female and 14 male patients. Ratio of female and male was 3.7:1.0. The median age onset of SLE was 11 years[3,16], whereas the median age onset of NPSLE was 13 years[6,18]. The major clinical presentations at the time of SLE diagnose were shown in Table 1. Renal, mucocutaneous and hematologic involvement were the major initial presentations noted in 51 (77.3%), 47 (71.2%), and 46 (69.7%) patients, respectively. Fifty-one patients (77.3%) had lupus nephritis and 46 patients underwent renal biopsies. Renal pathology was classified according to WHO classification of lupus nephritis. Most renal biopsy results were lupus nephritis class IV (63%). Mucocutaneous involvements presented as malar rash in 33 patients (50%), oral ulcer in 28 patients (42%), photosensitivity in 13 patients (19.7%), discoid rash in nine patients (13.6%), and dermal vasculitis in one patient (1.5%). Hematologic involvement presented as anemia (Hb <10 g/dl, Hct <30%) in 39 patients (59%), leukopenia (WBC <4,000 cell/mm^3) in 10 patients (15.1%), and thrombocytopenia (platelet <100,000 cell/mm^3) in seven patients (10.6%).

At the diagnosis of pediatric SLE, 76.7% (46/60 patients) had low C3 level and 56.9% (33/58 patients) had low C4 level. The median of C3 and C4 level were 0.3 (0.03 to 1.74) g/dL, and 0.08 (0.01 to 7.4) g/dL, respectively. ESR was performed in 59 patients and 55 patients (93.2%) had high ESR level (>20 mm/hour). The median ESR was 90 (10 to 140) mm/hour. Sixty-two and 48 patients had antinuclear antibodies (ANA) and anti-dsDNA measured, respectively. Sixty patients (96.8%) had positive ANA test and 39 patients (81.3%) had positive result of anti-dsDNA test. One of 30 patients (20%) had positive aPL, whereas eight of 17 patients (47.1%) had positive aCL.

Eight patients (12.1%) had NP manifestations on initial diagnosis of SLE, which were classified as seven seizure disorders and one cerebrovascular accident (CVA). Ten patients (58.8%) developed NP symptoms within the first year after diagnosis of SLE, while seven patients (41.2%) developed these symptoms one year later. The median duration of developed NP symptoms was 40 months.

In our study, the prevalence of NP manifestations was 25.8%. All of the patients had CNS manifestation (Table 2). No peripheral nervous system manifestations were observed. The most common NP manifestations were seizure (52.9%), CVA (29.4%), and movement disorder (17.6%), respectively. Other NP manifestation were headache (11.8%) and mood disorder (5.9%). Three patients (17.6%) had more than one manifestation (one seizure and one CVA).

Seizure was reported in nine patients (52.9%). Seven patients (77.8%) had generalized tonic-clonic

Table 1. Clinical features of SLE patients (n = 66)

Clinical features	n (%)
Male:female	14:52
Age of SLE diagnosed (years)*	11 (3, 16)
Initial presentation of SLE	
Mucocutaneous	47 (71.2)
Renal	51 (77.3)
Arthritis	24 (36.4)
Serositis	13 (19.7)
Neurologic disorders	8 (12.1)
Hematologic disorders	46 (69.7)
C3 level (g/L)*	0.31 (0.03, 1.74)
Low C3	46/60 (76.7)
C4 level (g/L)*	0.08 (0.01, 7.4)
Low C4	33/58 (56.9)
ESR*	90 (10, 140)
ANA positive	60/62 (96.8)
dsDNA positive	39/48 (81.3)
Antiphospholipid Ab positive	1/5 (20.0)
Anticardiolipin Ab positive	8/17 (47.1)
Renal biopsy	46/66 (69.7)
Class 2	8 (17.4)
Class 3	2 (4.3)
Class 4	29 (63.0)
Class 5	7 (15.2)

SLE = systemic lupus erythematosus; ESR = erythrocyte sedimentation rate; ANA = antinuclear antibodies; Ab = antibody
* Median (range)

seizure and two patients (22.2%) had focal-onset seizure. Recurrent episodes occurred in one patient, however, it was well controlled with standard anticonvulsant therapy. Five patients (29.4%) had CVA. We classified the patients as four ischemic strokes (80%) and one cerebral vein thrombosis (20%) with protein C deficiency and elevated aPL. Two patients (40%) had CVA occurred alone while CVA combined with seizure occurred in three patients (60%). No one had recurrent CVA. Chorea was observed in three patients (17.6%). All of them developed unilateral chorea after one year of SLE diagnosis. No recurrent episode of chorea was noted.

The prevalence of headache (11.8%) in the current study was lower than previous studies, which exhibited headache as the most common NPSLE manifestation[10,18,20,33]. The headache was classified into non-specific headache, which responded well to incrementation of oral prednisolone. Depression based on neuropsychological testing was noted in one patient (5.9%) during inactive phase of SLE. The symptom resolved after antidepressant therapy.

Specific investigations of NPSLE patients were shown in Table 3. CSF analysis was obtained in five patients. One patient (20%) had elevated CSF protein. The EEG was performed and revealed abnormality in 50% of cases. CT and MRI abnormalities were observed in 77.8% and 66.7%, respectively. Three, two, and two patients had infarction, brain atrophy, and subdural effusion on CT brain. The brain MRI abnormalities included three vasculitis and one infarction.

We compared the clinical and laboratory features between NPSLE and non-NPSLE patients (Table 4). There was no statistically significance in male gender and age of SLE diagnosis between groups. C4 levels of non-NPSLE patients was significantly lower than those of NPSLE groups. The C3 levels, ESR were not different between groups. The prevalence of positive ANA, anti-dsDNA antibodies, aPL, and aCL were not different between groups.

The treatment of NPSLE patients was shown in Table 5. All of the patients with NPSLE received oral corticosteroid as maintenance therapy. Additional treatments depended on severity of NP manifestations and the presence of thrombotic events. In mild NPSLE, one patient (5.9%) with depression received symptomatic treatment only. Two patients (11.8%) with non-specific headache responded well to oral prednisolone. In severe NPSLE, three patients (17.6%) were given pulse cyclophosphamide plus oral corticosteroid. One of them had evidence of cerebral vein thrombosis with protein c deficiency and demonstrated high titer of aPL, requiring warfarin and fresh frozen plasma administrations. The other

Table 2. Neuropsychiatric manifestations in 17 of 66 SLE patients (25.8%)

Manifestations	n (%)*
Cerebrovascular accident (CVA)	5 (29.4)
Seizure	9 (52.9)
Mood disorders	1 (5.9)
Movement disorders	3 (17.6)
Headache	2 (11.8)

* Some patients had multiple manifestations

Table 3. Investigations in 17 neuropsychiatric patients with SLE

Investigations	Abnormalities, n (%)	Abnormal finding
CSF (5)	1 (20.0)	High protein 1
EEG (2)	1 (50.0)	Slow wave 1
CT scan (9)	7 (77.8)	Infarction 3 Atrophy 2 Subdural effusion 2
MRI (6)	4 (66.7)	Infarction 1 Vasculitis 3

CSF = cerebrospinal fluid; EEG = electroencephalography; CT = computed tomography; MRI = magnetic resonance imaging

Table 4. Comparison of clinical features at the time of diagnosis between patients with NPSLE (n = 17) and non-NPSLE (n = 49)

Clinical features	NPSLE n (%)	Non-NPSLE n (%)	*p*-value
Male gender	3 (17.6)	11 (22.4)	1.00
Low C3 Level (g/L)	12/16 (75.0)	34/44 (77.3)	1.00
Low C4 Level (g/L)	6/15 (40.0)	27/43 (62.8)	0.14
ESR*	76.5 (18, 125)	95 (10, 140)	0.18
ANA positive	16/16 (100)	44/46 (95.7)	1.00
dsDNA positive	7/11 (63.6)	32/37 (86.5)	0.18
Antiphospholipid positive	0/2 (0)	1/3 (33.3)	1.00
Anticardiolipin positive	4/7 (57.1)	4/10 (40.0)	0.64

NPSLE = SLE patients with neuropsychiatric disorders
* Median (range)

Table 5. Treatment of 17 patients with NPSLE

Medications	n (%)
Prednisolone	2 (11.8)
Prednisolone + cyclophosphamide	3 (17.6)
Prednisolone + methylprednisolone	11 (64.7)
Prednisolone + cyclophosphamide + hydroxychloroquine	1 (5.9)

NPSLE = SLE patients with neuropsychiatric disorders

11 patients (6 seizure, 4 CVA, 1 chorea) were given intravenous pulse methylprednisolone (30 mg/kg/day or 1 gm daily) for three days and oral prednisolone. One patient (5.9%) with chorea and active malar rash received a combination of pulse cyclophosphamide, hydroxychloroquine, and corticosteroid. All patients with CVA received low dose aspirin (3 to 5 mg/kg/day) for stroke prevention.

Discussion

The prevalence of pediatric NPSLE (25.8%) in the current study was comparable to previous studies, ranging from 22% to 95%[5,8,10,11,13,16,19]. The median age onset of NPSLE in our study was similar to the previous study[34]. The prevalence of multiple NP manifestations in our report were similar to the previous study[35]. The present study demonstrated seizure and CVA as the most common NPSLE manifestations which were similar to reports from Supavekin et al[12], Yu et al[11], and Mok et al[19]. Besides, seizure was similarly reported as a single attack of generalized tonic-clonic in nature and usually well controlled with antiepileptic drug monotherapy[11,20]. In contrast, headache, mood disorders, and psychosis were more frequently described in reports from western countries[10,18]. We hypothesized that Ethnicity (Asian versus non-Asian) may be one of the potential predictors of NP manifestations and Asian pediatric SLE have higher risk of developing seizure than non-Asian population.

We observed headache in only 11.8% of NPSLE patients. The lower prevalence of headache compared to other studies is probably due to rigorous criteria of diagnosis. We excluded headache from secondary causes such as aseptic meningitis, drug-induced pseudotumor cerebri, tumors, and other brain lesions. Moreover, the occurrence of headache might be underreported in medical records.

Although previous studies exhibited the correlation of aPLs antibodies with severe NPSLE such as CVA, seizure, and chorea[18,22-24], we were not able to demonstrate such findings. Apparently, these tests were performed in a small number of SLE patients in the present study. Regarding specific investigations, CSF analysis, EEG, CT scan, and MRI findings were non-specific to NP manifestations. Similar to the previous studies[17,20], vasculitis was a common abnormality on MRI in NPSLE patients.

Concerning the treatment of NPSLE, previous studies noted that corticosteroids had benefit in treating migraine headache[25,26]. Other types of headache had no mention about corticosteroid treatment. We proposed a role of corticosteroid for treatment of non-specific headache in SLE patients. In severe NPSLE (seizure, CVA, chorea), the ideal therapeutic regimen is still controversial between cyclophosphamide and methyl-prednisolone due to absence of randomized controlled trials comparing the effectiveness and safety[21,27-31]. The optimal treatment in severe NPSLE continues to be based on expert opinion and clinical experience[33]. In our small series, SLE patients received pulse cyclophosphamide, pulse methylprednisolone, and oral prednisolone. Thus, we are not able to clarify the response of NPSLE to any specific therapy.

Our study had some limitations including small sample size and validity of retrospective review. Symptoms of mild degree, duration of follow-up, and outcome were underreported. Therefore, the prevalence of NPSLE manifestations in the present study may be underestimated.

In conclusion, the prevalence of NP manifestations was approximately one-fourth of pediatric SLE patients. Most of them exhibited NP symptoms within the first year after diagnosis. More than 50% of NPSLE patients had abnormal findings on brain imaging. Interestingly, the prevalence of seizures in Asian pediatric SLE was found to be more significant than non-Asian population. We hypothesized that ethnicity may be one of the potential predictors of NP manifestations. Finally, large prospective studies are warranted to identify the accurate prevalence, risk factors, and proper management of NPSLE patients.

What is already known on this topic?

Previous literatures showed the prevalence of pediatric NPSLE varied from a low of 20% to a high of 95%, and no previous studies identified characteristics of pediatric NPSLE in Thailand.

What this study adds?

This study found the prevalence of pediatric NPSLE was 25.8%, and added the characteristic of NPSLE. The most common symptoms were seizure (52.9%), stroke (29.4%), and movement disorder (17.6%). Interestingly, the prevalence of seizures in Asian pediatric SLE was found to be more significant than non-Asian population. Ethnicity may be one of the potential predictors of NP manifestations.

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Potential conflicts of interest

None.

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อาการแสดงทางระบบประสาทในผู้ป่วยเด็กเอสแอลอี

ภิรดี สุวรรณภักดี, ชาครินทร์ ณ บางช้าง, อดิสรณ์ ลำเพาพงศ์

ภูมิหลัง: *อาการแสดงทางระบบประสาทในผู้ป่วยเด็กเอสแอลอีพบได้ไม่บ่อย*

วัตถุประสงค์: *เพื่อศึกษาอุบัติการณ์ และอาการแสดงของผู้ป่วยเด็กเอสแอลอีที่มีอาการแสดงทางระบบประสาท*

วัสดุและวิธีการ: *ทบทวนเวชระเบียนผู้ป่วยเด็กเอสแอลอี ที่ได้รับการวินิจฉัย ตั้งแต่เดือนมกราคม พ.ศ. 2531 ถึง สิงหาคม พ.ศ. 2552 พบผู้ป่วยเด็กเอสแอลอีจำนวน 66 ราย หากผู้ป่วยมีอาการคล้ายเอสแอลอี (lupus-like symptom) จากสาเหตุอื่น จะคัดออกจากการศึกษา*

ผลการศึกษา: *พบผู้ป่วยเด็กเอสแอลอีที่มีอาการแสดงทางระบบประสาทจำนวน 17 ราย (25.8%) โดยมีอายุเฉลี่ยเท่ากับ 13 ปี (6, 18 ปี) ผู้ป่วยจำนวน 8 ราย (12.1%) มีอาการแสดงทางระบบประสาท ตั้งแต่เริ่มการวินิจฉัยเอสแอลอี ผู้ป่วย 10 ราย (58.8%) มีอาการแสดงภายในหนึ่งปีแรกหลังการวินิจฉัยโรค ในขณะที่ผู้ป่วย 7 ราย (41.2%) มีอาการแสดงภายหลัง 1 ปีหลังการวินิจฉัยโรค อาการแสดงทางระบบประสาทที่พบบ่อย 3 อันดับแรก ได้แก่ ชัก (52.9%) สมองขาดเลือดเฉียบพลัน (29.4%) และการเคลื่อนไหวผิดปกติ (17.6%) ไม่พบว่าผลเลือดทางห้องปฏิบัติการใด ๆ สัมพันธ์กับอาการแสดงทางระบบประสาท ส่วนภาพถ่ายคอมพิวเตอร์สมองและคลื่นแม่เหล็กไฟฟ้าสมองจะพบความผิดปกติในผู้ป่วยที่มีอาการแสดงทางระบบประสาทมากกว่า 50% ได้แก่ สมองขาดเลือด หลอดเลือดอักเสบ สมองฝ่อ และมีน้ำคั่งใต้ชั้นดูรา (subdural effusion)*

สรุป: *การศึกษานี้พบว่าผู้ป่วยเอสแอลอีที่มีอาการแสดงทางระบบประสาทส่วนใหญ่จะมีอาการภายใน 1 ปีหลังจากการวินิจฉัยโรค และเป็นที่น่าสนใจว่า พบอุบัติการณ์ของอาการชักในเด็กไทยสูงกว่าการศึกษาในต่างประเทศนอกเหนือทวีปเอเชีย อาจเป็นไปได้ว่าเชื้อชาติเป็นปัจจัยหนึ่งที่มีผลต่ออาการแสดงทางระบบประสาทในผู้ป่วยเด็กเอสแอลอี*