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Case Report

Focal Digital Clubbing as an Initial Symptom of Adolescent-Onset Systemic Lupus Erythematosus: A Case Report

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Abstract

Digital clubbing is typically linked to pulmonary or cardiovascular diseases. There have been no reported cases of systemic lupus erythematosus (SLE) where the initial complaint was solely focal digital clubbing and a chilblain-like rash on the fingers. A 13-year-old girl experienced digital clubbing for 3 months. Physical examination revealed focal digital clubbing of the thumb, index finger, and middle finger on her right hand, along with slight blotchy erythema assessed as a chilblain-like rash. Laboratory data showed hypocomplementemia and a high anti-double-stranded DNA antibody titer. She was also positive for antiphospholipid antibodies, had prolonged prothrombin time and activated partial thromboplastin time, and elevated anti-prothrombin antibody levels. She was diagnosed with SLE complicated by hypoprothrombinemia associated with lupus anticoagulant. Prednisolone treatment resolved the digital clubbing and laboratory abnormalities. Digital clubbing may be an initial symptom of SLE. Screening for autoimmune diseases should be considered when a patient presents with digital clubbing not associated with pulmonary or cardiovascular diseases or malignant tumors.

Keywords: Antiphospholipid antibodies, aPLs, digital clubbing, hypoprothrombinemia, SLE, systemic lupus erythematosus

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease that can affect various organs and tissues. In children, the clinical course of SLE is both more acute and more severe than in adults.[1] A Japanese surveillance study revealed that the initial symptoms of pediatric SLE are primarily fever, malar rash, joint symptoms, and abnormal urine.[2] Digital clubbing has rarely been reported as a manifestation of SLE.[3,4] Moreover, there have been no reported cases of SLE in which digital clubbing and a chilblain-like rash on the fingers were the sole presenting symptoms.

Herein, an SLE patient diagnosed on the basis of these unusual clinical presentations is presented and associations between SLE and digital clubbing are considered.

Case Presentation

A 13-year-old girl who had been experiencing digital clubbing for 3 months was referred to a primary care doctor. Her past medical history was insignificant, and menarche had already been established at age 11. Family history was notable for her maternal aunt with an autoimmune disease, for which the details were unclear. As the screening examination showed signs of inflammation and anti-nuclear antibody (ANA) positivity, she was referred to the hospital for further examination.

A physical examination revealed focal digital clubbing of the thumb, index finger, and middle finger of her right hand with a positive Schamroth sign, the obliteration of the diamond-shaped window normally produced between the proximal tips of the nails when the distal phalanges are opposed,[5] while the other fingers and toes did not show any abnormalities (Figure 1A). She was also evaluated by a dermatologist; thermography showed decreased blood flow consistent with digital clubbing (Figure 2), and there was slight blotchy erythema on her thumb that was assessed as a chilblain-like rash (Figure 3).

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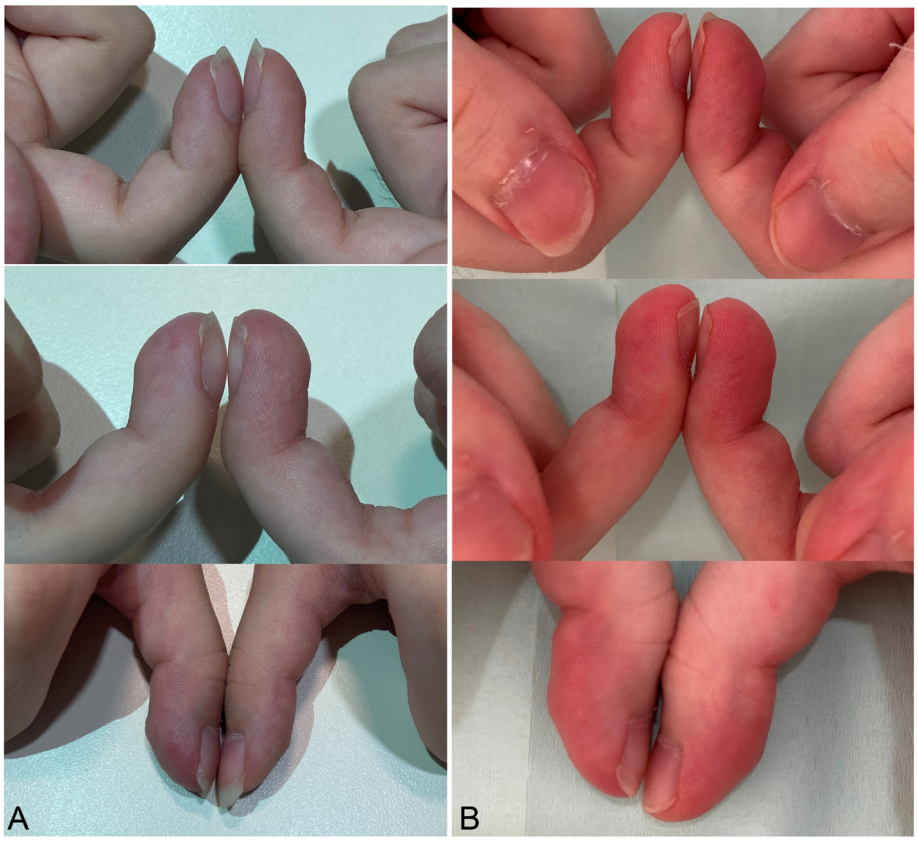

Figure 1. (A) The patient's fingers upon admission. Focal digital clubbing was observed in the thumb and index finger. The Schamroth sign was positive in the affected fingers. (B) The patient's fingers at discharge. Digital clubbing was almost inconspicuous.

Her laboratory data showed hypocomplementemia, a high anti-double-stranded DNA (anti-ds-DNA) antibody titer, and positivity for ANA (Table S1). Her urinalysis did not show hematuria or proteinuria. No organ complications of SLE, such as those affecting the lungs, bowel, heart, and brain, were observed through imaging tests.

Systemic lupus erythematosus was suspected based on laboratory results; however, the patient did not satisfy the American College of Rheumatology (ACR) classification criteria for SLE.[6] On the other hand, she did meet the Systemic Lupus International Collaborating Clinics[7] criteria as she had a chilblain-like rash indicative of chronic cutaneous lupus. She also showed antiphospholipid antibody (aPL) positivity, along with prolongation of both PT (prothrombin time/international normalized ratio: 1.61, reference interval: 0.80-1.20) and aPTT (activated partial thromboplastin time: 83.2 seconds, reference interval: 27.0-39.9). Evaluation of coagulation factors revealed decreased factor II activity (30%, reference interval: 75-135). Further examination showed a significantly elevated anti-prothrombin antibody level of greater than 200 U/mL (reference interval: <30). Thus, she was diagnosed with SLE and concomitant hypoprothrombinemia associated with lupus anticoagulant.

She was administered 60 mg of prednisolone (PSL) daily. Fourteen days later, PT and aPTT were normalized. Although there were no urinary abnormalities, it was decided to perform a kidney biopsy due to concern for silent lupus nephritis, which refers to histopathological evidence of nephritis in SLE patients without overt clinical or biochemical signs of renal involvement. The kidney biopsy was performed on day 19 after the initiation of PSL treatment, which revealed International Society of Nephrology/Renal Pathology Society class II lupus nephritis. The anti-ds-DNA antibody and C3 titers were normalized on day 21. Additional treatment with mizoribine and hydroxychloroquine was initiated on day 28. Subsequently, PSL was successfully tapered from day 34 while maintaining remission. She was discharged 2 months after admission. Her digital clubbing was almost inconspicuous at discharge (Figure 1B), and did not show relapse during the subsequent 6 months. Written informed consent for the article was obtained from the patient and parents of the patient.

Main Points

- This is the first report of a systemic lupus erythematosus (SLE) patient whose main complaint was limited to digital clubbing of and a chilblain-like rash on the fingers.
- Digital clubbing may be an initial symptom of SLE.
- Screening tests for autoimmune disease should be considered when a patient presents with digital clubbing uncomplicated by pulmonary disease, cardiovascular disease, or malignant tumors.

Discussion

We here present a female case of adolescent-onset SLE diagnosed from the unusual initial symptoms of solely digital clubbing and a chilblain-like rash on the fingers. She also had also coagulation abnormalities, including aPL positivity. She had not only prolonged aPTT but also prolonged PT. Further examination showed low factor II activity and anti-prothrombin antibody positivity. Thus, the authors assessed her with hypoprothrombinemia associated with lupus anticoagulant.

The prevalence of aPLs is approximately 40% in patients with pediatric SLE.[8] These patients are susceptible to thrombosis, representing antiphospholipid antibody syndrome (APS). These aPLs are associated with aPTT prolongation. On the other hand, the presence of factor II deficiency along with LA positivity is typical of lupus anticoagulant hypoprothrombinemia syndrome (LAHPS), which shows both aPTT and PT prolongation. Lupus anticoagulant hypoprothrombinemia syndrome is a rare manifestation of SLE, presenting with bleeding.[9] Although the patient did not exhibit overt bleeding and the clinical picture did not fully align with typical cases, an LAHPS-like condition was suspected. Therefore, treatment was initiated prior to the kidney biopsy, given the potential risk of bleeding.

Acquired digital clubbing is most often associated with pulmonary or cardiovascular diseases.[10,11] Although the mechanism of digital clubbing is not fully understood, it has been suggested that vascular endothelial growth factor (VEGF), a platelet-derived growth factor that is induced by hypoxia, plays a central role.[11] Generally, digital clubbing due to systemic hypoxic conditions occurs on all fingers; however, in this patient, it occurred partially. From this point of view, it was suspected that susceptibility to thrombosis due to SLE itself or

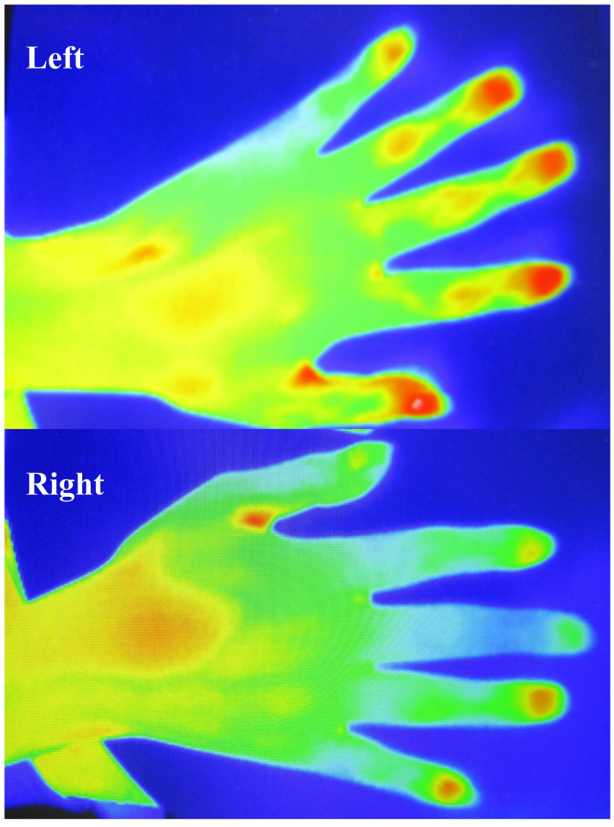

Figure 2. The findings of thermography. Decreased blood flow was observed, consistent with digital clubbing.

as a complication of aPLs in this patient may have led to the formation of microthrombi, resulting in focal tissue hypoxia of the fingertips and the formation of focal digital clubbing. This hypothesis appears to be supported by the hand thermography findings, which showed focal decreased blood flow concurrent with the digital clubbing.

Mackie[3] reported a 50-year-old female who presented with finger clubbing accompanied by intolerance to sunlight and facial erythema, and who was diagnosed with SLE. This patient was suspected to have hypoxia of finger pulp to some degree. Harris et al[4] presented the case of a 48-year-old male diagnosed with APS, not SLE, who presented with digital clubbing. The authors surmised that platelet aggregation and microthrombi formation were caused by anti-cardiolipin antibodies and resulted in the release of platelet-derived growth factor, leading to increased capillary permeability and connective tissue hypertrophy.

This patient did not meet the ACR criteria for the diagnosis of SLE due to the absence of typical symptoms. As she was screened at the time of onset of her initial limited symptoms, such as digital clubbing and a chilblain-like rash on the fingers, one could diagnose SLE before progression to organ damage. It is suggested that when a patient complains of digital clubbing, screening tests for SLE or aPLs should be considered.

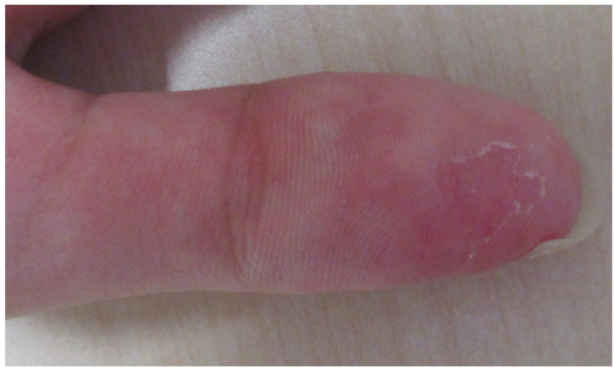

Figure 3. The patient's right thumb showed blotchy erythema on the thumb, which a dermatologist assessed as a chilblain-like rash.

This report has several limitations. First, the mechanism underlying finger clubbing remains a hypothesis as a biopsy of the fingertip skin could not be performed and serum VEGF levels could not be measured. However, the thermography findings strongly suggested decreased blood flow in the clubbed fingers, which is indicative of tissue hypoxia. Second, assuming the hypothesis that focal tissue hypoxia leads to digital clubbing, it could not be determined whether it was attributable to SLE itself or aPLs. It remains unknown whether digital clubbing occurs in SLE patients without complications associated with aPLs. Future studies are required to accumulate similar cases and to confirm the hypothesis.

In conclusion, while there have been few reports presenting digital clubbing concomitant with other symptoms of autoimmune disease, this is the first report of an SLE patient whose main complaint was limited to digital clubbing of and a chilblain-like rash on the fingers. It is suggested that digital clubbing may be an initial symptom of SLE. Screening tests for autoimmune disease should be considered when a patient presents with digital clubbing uncomplicated by pulmonary or cardiovascular disease.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Written informed consent was obtained from the patient and parents of the patient who agreed to take part in the study.

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Supplementary Table 1 . Laboratory Findings

		(reference interval)			(reference interval)
WBC	5,740 /mm^3	(3380-8600)	Albumin	4.3 g/dL	(4.1-5.1)
RBC	4.21 x 10^6 mm^3	(3.86-4.92)	Creatinine	0.49 mg/dL	(0.46-0.79)
Hgb	9.4 g/dL	(11.6-14.8)	IgG	1814 mg/dL	(861-1747)
Platelet	285 x 10^3 mm^3	(158-348)	CH50	< 10.0 U/mL	(30.0-45.0)
			C3	69.9 mg/dL	(73.0-138.0)
PT/INR	1.61	(0.80-1.20)	C4	2.2 mg/dL	(11.0-31.0)
aPTT	83.2 Second	(27.0-39.9)	ESR	29 mm/h	(3-15)
D-dimer	< 0.50 μg/mL	(0-0.50)	RF	< 3 IU/mL	(<15.0)
			anti-ds-DNA antibody	42.3 IU/mL	(< 12.0)
factor II	30 %	(75-135)	anti-Sm antibody	4.8 U/mL	(< 7.0)
factor V	68 %	(27.0-39.9)	anti-CL-β2GPI antibody	7.9 U/mL	(< 3.5)
factor X	84 %	(70-130)	anti-RNP antibody	0.7 U/mL	(< 3.5)
PIVKA-II	13 mAU/mL	(< 40)	anti-SS-A/Ro antibody	< 0.4 U/mL	(< 7.0)
			anti-SS-B/La antibody	0.4 U/mL	(< 7.0)
LA (diluted Russel's viper venom time)	3.1	(< 1.2)	ANA (fluorescent antibody method)	1:160	(< 1:40)

RBC, red blood cell; PT/INR, prothrombin time/international normalized ratio; aPTT, activated partial thromboplastin time, PIVKA-II; protein induced by vitamin K absence or antagonist II, LA; lupus anticoagulant, ESR; erythrocyte sedimentation rate, RF; rheumatoid factor, ant-ds-DNA antibody; anti-double-stranded DNA antibody, anti-CL- 2GPI antibody; anti-cardiolipin and beta 2 glycoprotein I antibody, anti-RNP antibody; anti-ribonucleoprotein antibody, ANA; anti-nuclear antibody

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