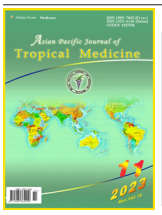

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Association between the Lung Immune Prognostic Index and mortality in patients with idiopathic inflammatory myopathy−associated interstitial lung disease

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ABSTRACT

Objective: To explore the association between the Lung Immune Prognostic Index (LIPI) and 1-year all-cause mortality in patients with idiopathic inflammatory myopathy related interstitial lung disease (IIM-ILD).

Methods: Patients who were diagnosed with IIM-ILD at West China Hospital, Sichuan University from January 2008 to December 2021 were retrospectively included and categorized into three groups based on LIPI. Univariable and multivariable Cox proportional hazards models were conducted to explore potential association between the LIPI and patients' mortality.

Results: A total of 1 116 patients were screened, and 830 were included in this study. The multivariable Cox analysis showed that, compared with patients with poor LIPI, the hazard ratio (*HR*) for all-cause 1-year mortality was 0.22 (95% *CI* 0.05-0.93, *P*=0.04) for patients in the good LIPI group (LDH<250 IU/L and dNLR<3). After excluding patients lost to follow-up within one year, a similar result was found for LIPI (*HR* 0.20, 95% *CI* 0.05-0.86; *P*=0.03).

Conclusions: Good LIPI was independently associated with decreased risk of all-cause 1-year mortality in patients with IIM-ILD. This easy-to-obtain index might be served as a potential marker for assessing the prognosis of IIM-ILD.

KEYWORDS: Idiopathic inflammatory myopathy; Interstitial lung disease; Lung Immune Prognostic index; All-cause mortality

1. Introduction

The idiopathic inflammatory myopathy associated interstitial lung disease (IIM-ILD) is the most prevalent extra-muscle manifestation of idiopathic inflammatory myopathy (IIM), which is a heterogenous group of diseases with various subtypes, clinical manifestations,

Significance

The Lung Immune Prognostic Index is a marker of pulmonary immunity derived from lactate dehydrogenase and peripheral blood cells. The association between this index and mortality risk of patients with idiopathic inflammatory myopathy related interstitial lung disease is unknown. This retrospective cohort study reports that good Lung Immune Prognostic Index (LDH<250 IU/L and dNLR<3) independently decreases the patients' risk of all-cause mortality in 1-year by around 88% indicating that it can be considered as a potential biomarker for assessing patients' prognosis.

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treatment responses and prognosis[1-3]. To delay the progression of lung disease, interventions should be commenced early and personally. However, effective and personalized interventions require comprehensive evaluation of disease severity, acute exacerbation risk and mortality risk. Thus, it is imperative to develop user-friendly markers for IIM-ILD that can be applied in routine clinical settings.

Lung Immune Prognostic Index (LIPI) is a marker of pulmonary immunity derived from lactate dehydrogenase (LDH) and peripheral blood cells[4]. Serum LIPI elevations have shown prognostic values for mortality in many diseases, including lung cancers[5-8]. But the effects of LIPI level in patients with IIM-ILD remain poorly understood. Our previous studies showed that creatase was associated with long-term and short-term mortality for patients with IIM-ILD[9], and systemic inflammation and immunity index was positively associated with respiratory failure risk during hospitalization. That implies LIPI, a combination of creatase and blood-derived immunity, might be served as a potential marker for assessing the severity and prognosis of IIM-ILD.

In this study, we will explore the associations between LIPI and all-cause mortality risk among patients with IIM-ILD. Within a large hospital-based cohort of adults, we demonstrated that poor LIPI would be associated with higher risk of mortality. LIPI level could be a potential biomarker for assessing the prognosis of IIM-ILD. This findings will provide valuable insights for guiding the treatment of IIM-ILD.

2. Subjects and methods

2.1. Study population

This retrospective hospital-based cohort study was approved by the Institutional Review Board of West China Hospital, Sichuan University [approval No. 2022(1996)], and informed consent was waived. Patients diagnosed with IIM-ILD at West China Hospital from January 2008 to December 2021 were included for analysis. IIM was diagnosed according to the Bohan & Peter Diagnostic Criteria[10] or the 2004 European Neuromuscular Centre criteria[11], and interstitial lung disease (ILD) was identified by chest high-resolution computed tomography (HRCT). In this study, diseases including clinically diagnosed amyopathic dermatomyositis (ADM), antisynthetase syndrome (ASS), polymyositis (PM), dermatomyositis (DM), immune-mediated necrotizing myopathy (IMNM) and nonspecific myositis (NSM) were all refered as IIM, while overlap myositis and cancer associated myositis were excluded, patients admitted to the intensive care unit initially were excluded, and patients without a baseline LIPI were also excluded.

2.2. Assessment of LIPI

Peripheral venous blood samples were collected and sent to clinical laboratory for further measurement. For each patient, the blood examination results conducted closest to the date of enrollment within 3 days, were analyzed. LIPI was categorized into three groups by LDH and neutrophils/(leukocytes minus neutrophils) ratio (dNLR): good LIPI (LDH<250 IU/L and dNLR<3), middle LIPI (LDH 250 IU/L or dNLR≥3), and poor LIPI (LDH 250 IU/L and dNLR≥3).

2.3. Radiological evaluation

Chest HRCT images and videos were retrospectively collected and reviewed by two experienced respiratory physicians (Dr. Zhu and Dr. He) and a radiologist (Dr. Yan) who were blinded to the clinical information. HRCT scan results were categorized as definite usual interstitial pneumonia (UIP), probable UIP, possible UIP or impossible UIP pattern[12,13]. For each patient, the HRCT examination results which were conducted near the time of enrollment within 2 months, were analyzed. HRCT scan results with poor quality were excluded.

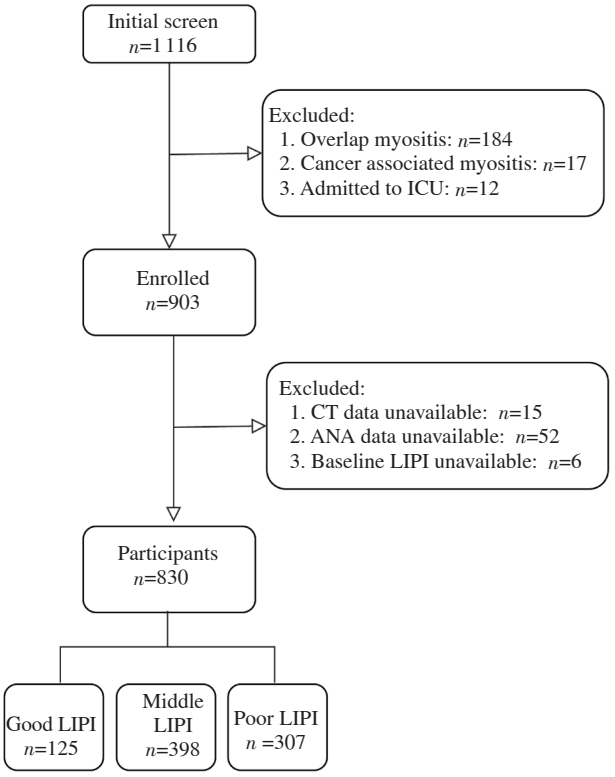

Figure 1. Flow chart of patient inclusion and grouping. ICU: intensive care unit; CT: computed tomography; ANA: antinuclear antibody; LIPI: Lung Immune Prognostic Index.

2.4. Follow–up

Follow-up started on the day of admission and ended at the occurrence of death, lost to follow-up, or June 2022, whichever occurred first. Patients were reviewed by clinical visits or phone reviews depending on the disease progression. Death caused by any causes were retrieveded from electrical medical records, or by telephone check with patients' family members. Patients who were loss of follow-up were assumed alive. The primary outcome of this study was all-cause 1-year mortality.

2.5. Statistical analyses

Data of baseline characteristics are expressed as mean ± standard deviation or median (interquartile range) for continuous variables, and percentages for categorical variables, differences among the three LIPI groups were analyzed with ANOVA tests, *Chi*-squared tests or Fisher's exact tests where appropriate.

Kaplan–Meier curve was plotted, and log-rank test was used to compare mortality rates among the three groups. Multivariable Cox proportional hazards model was then used to calculate hazard ratio (*HR*) for the association between LIPI and all-cause 1-year mortality, with the poor LIPI group as the reference group. Cofounders were adjusted, which were selected according to previous studies and clinical experience. Albumin (ALB) was categorized as normal group (≥40 g/L) and abnormal group(<40 g/L) before entering the model. R software (version 4.1.2) was used for all statistical analysis with a two-tailed *P* value of less than 0.05 as statistical significance.

3. Results

3.1. Baseline characteristics of study participants

A total of 1 116 patients were initially screened, and 830 patients were enrolled (Figure 1). Average age of the participants were (51.1±11.8) years and 439 (52.9%) were elder than 50 years old. 555 (66.9%) Patients were female, 657 (79.2%) were never smokers, and 331 (39.9%) were newly diagnosed patients. There were 72 (8.7%) ADM, 91 (11.0%) ASS, 450 (54.2%) DM, 11 (1.3%) IMNM, 108 (13.0%) NSM and 98 (11.8%) PM patients in our cohort, respectively. Table 1 presents the baseline characteristics of patients grouped by LIPI.

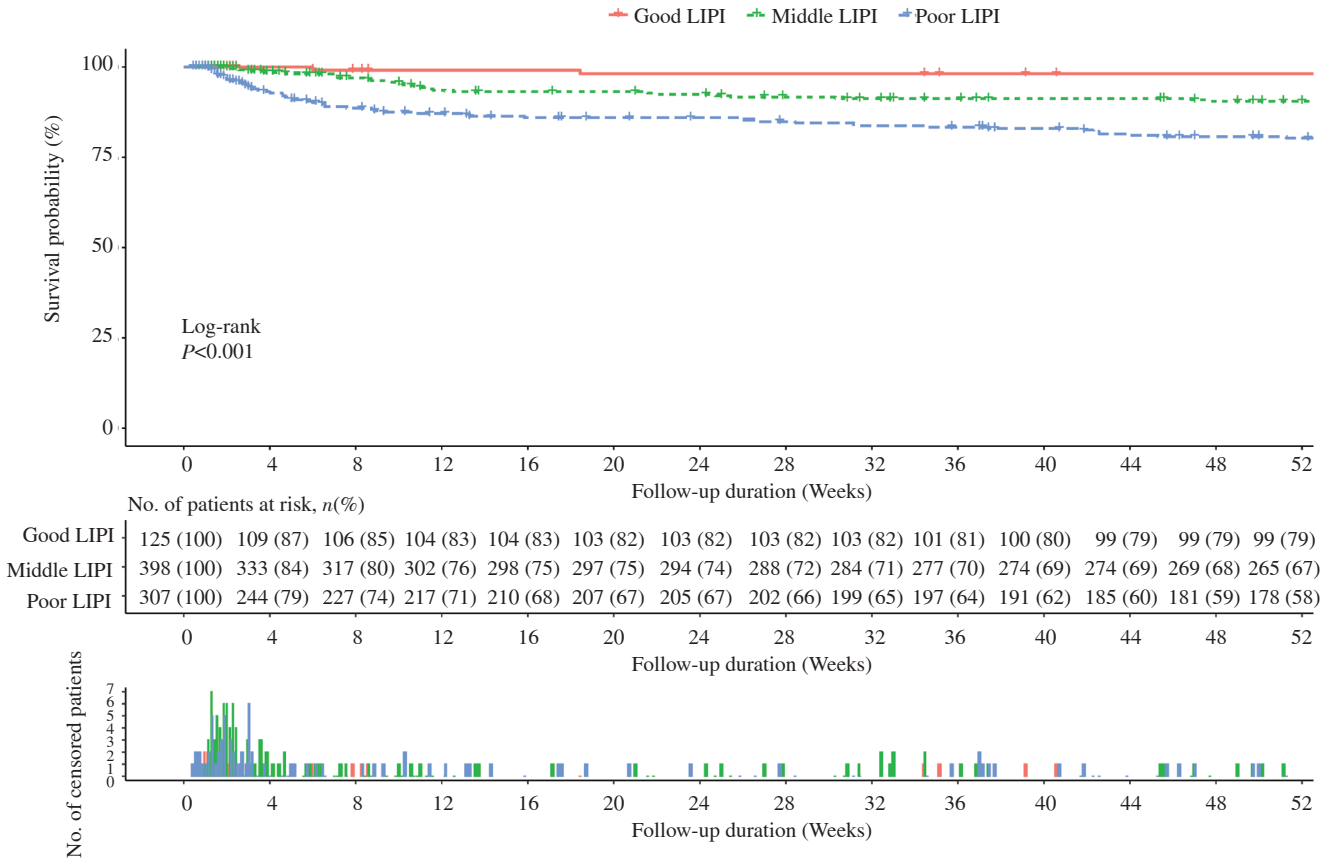

Figure 2. Kaplan–Meier survival curves of patients with IIM-ILD grouped by LIPI. LIPI: Lung Immune prognostic index.

Table 1. Baseline clinical and laboratory data of the three groups [*n* (%)].

Parameters	Good LIPI (*n*=125)	Middle LIPI (*n*=398)	Poor LIPI (*n*=307)	Total cohort (*n*=830)	*F*/ χ^2	*P* values
Follow-up weeks*	168 (0.86, 605)	124 (1.00, 698)	86.3 (0.43, 636)	117 (0.43, 698)	-	-
Age, years#	50.4±11.6	51.3±12.1	51.1±11.5	51.1±11.8	0.281	0.755[a]
Age <50 years	61 (48.8)	195 (49.0)	135 (44.0)	391 (47.1)	1.923	0.382[b]
Age ≥50 years	64 (51.2)	203 (51.0)	172 (56.0)	439 (52.9)		
Sex						
Female	87 (69.6)	274 (68.8)	194 (63.2)	555 (66.9)	2.995	0.224[b]
Male	38 (30.4)	124 (31.2)	113 (36.8)	275 (33.1)		
Smoker						
No	101 (80.8)	323 (81.2)	233 (75.9)	657 (79.2)	3.147	0.207[b]
Yes	24 (19.2)	75 (18.8)	74 (24.1)	173 (20.8)		
Newly diagnosed						
No	70 (56.0)	224 (56.3)	205 (66.8)	499 (60.1)	9.002	0.011[b]
Yes	55 (44.0)	174 (43.7)	102 (33.2)	331 (39.9)		
CCI						
High	50 (40.0)	172 (43.2)	162 (52.8)	384 (46.3)	8.685	0.013[b]
Low	75 (60.0)	226 (56.8)	145 (47.2)	446 (53.7)		
Lab results#						
ALB	38.4±5.25	35.4±5.17	32.9±5.37	34.9±5.57	51.620	<0.001[a]
AST/ALT	1.19±0.46	1.45±2.10	1.67±4.57	1.49±3.14	1.079	0.340[a]
ANA						
Negative	59 (47.2)	171 (43.0)	150 (48.9)	380 (45.8)	2.546	0.280[b]
Positive	66 (52.8)	227 (57.0)	157 (51.1)	450 (54.2)		
CT patterns						
Non-UIP	115 (92.0)	374 (94.0)	292 (95.1)	781 (94.1)	1.573	0.456[b]
UIP	10 (8.0)	24 (6.0)	15 (4.9)	49 (5.9)		
Clinical diagnosis						
ADM	13 (10.4)	34 (8.5)	25 (8.1)	72 (8.7)	-	<0.001[c]
ASS	32 (25.6)	30 (7.5)	29 (9.4)	91 (11.0)		
DM	53 (42.4)	220 (55.3)	177 (57.7)	450 (54.2)		
IMNM	1 (0.8)	5 (1.3)	5 (1.6)	11 (1.3)		
NSM	11 (8.8)	55 (13.8)	42 (13.7)	108 (13.0)		
PM	15 (12.0)	54 (13.6)	29 (9.4)	98 (11.8)		
High dose corticoids						
No	94 (75.2)	230 (57.8)	151 (49.2)	475 (57.2)	24.658	<0.001[b]
Yes	31 (24.8)	168 (42.2)	156 (50.8)	355 (42.8)		
IVIG						
No	122 (97.6)	382 (96.0)	276 (89.9)	780 (94.0)	14.722	<0.001[b]
Yes	3 (2.4)	16 (4.0)	31 (10.1)	50 (6.0)		
Pulmonary infection						
0	79 (63.2)	183 (46.0)	71 (23.1)	333 (40.1)	109.640	<0.001[b]
1	45 (36.0)	166 (41.7)	138 (45.0)	349 (42.0)		
2	1 (0.8)	42 (10.6)	69 (22.5)	112 (13.5)		
3	0 (0.0)	7 (1.8)	29 (9.4)	36 (4.3)		
Respiratory failure						
No	125 (100.0)	385 (96.7)	252 (82.1)	762 (91.8)	62.580	<0.001[b]
Yes	0 (0.0)	13 (3.3)	55 (17.9)	68 (8.2)		
Death in one year						
No	123 (98.4)	367 (92.2)	256 (83.4)	746 (89.9)	26.579	<0.001[b]
Yes	2 (1.6)	31 (7.8)	51 (16.6)	84 (10.1)		
Lost to follow-up in one year	24 (19.2)	103 (25.9)	78 (25.4)	205 (24.7)		

ADM: clinically amyopathic dermatomyositis; ASS: antisynthetase syndrome; DM: dermatomyositis; IMNM: immune-mediated necrotizing myopathy; NSM: nonspecific myositis; PM: polymyositis; CCI: charlson comorbidity index; AST: aspartate transaminase; ALT: alanine transaminase; ANA: antinuclear antibody; IVIG: intravenous use of immunoglobulin; ALB: albumin; UIP: usual interstitial pneumonia (UIP pattern and Probable UIP pattern are included); CT: computed tomography; Bold indicates statistical significance. *Data were expressed as median (IQR); #Data were expressed as mean±SD; - statistical analysis not conducted; a: ANOVA test, b: Pearson's *Chi*-squared test, c: Fisher's exact test.

Table 2. Univariate and multivariate Cox regression analysis.

LIPI	*n*	Event, *n* (%)	Hazard Ratio (95% *CI*)					
			Unadjusted	*P*	Model 1[a]	*P*	Model 2[b]	*P*
Poor	307	51 (16.6)	Reference		Reference		Reference	
Middle	398	31 (7.8)	0.44 (0.28-0.68)	<0.001	0.66 (0.41-1.05)	0.079	0.75 (0.47-1.21)	0.239
Good	125	2 (1.6)	0.08 (0.02-0.35)	<0.001	0.15 (0.04-0.62)	0.009	0.22 (0.05-0.93)	0.040

a: model 1 was adjusted for respiratory failure; b: model 2 was adjusted for age, AST/ALT, ALB, ANA, CT pattern, pulmonary infection, respiratory failure, CCI and clinical subtypes.

Table 3. Univariate and multivariate Cox regression analysis excluding patients lost to follow-up.

LIPI	*n*	Event, *n* (%)	Hazard Ratio (95% *CI*)					
			Unadjusted	*P*	Model 1[a]	*P*	Model 2[b]	*P*
Poor	307	51 (16.6)	Reference		Reference		Reference	
Middle	398	31 (7.8)	0.44 (0.28-0.68)	<0.001	0.66 (0.41-1.05)	0.079	0.75 (0.47-1.21)	0.239
Good	125	2 (1.6)	0.08 (0.02-0.35)	<0.001	0.15 (0.04-0.62)	0.009	0.22 (0.05-0.93)	0.040

a: model 1 was adjusted for respiratory failure; b: model 2 was adjusted for age, AST/ALT, ALB, ANA, CT pattern, pulmonary infection, respiratory failure, CCI and clinical subtypes.

3.2. Association between LIPI and mortality in patients with IIM–ILD

As shown in Table 1, the number of newly diagnosed patients included in the poor LIPI group was more (*n*=307) than that in the good LIPI group (*n*=125), more patients in the poor LIPI group had high charlson comorbidity index (CCI), had received high doses of corticoid and intravenous use of immunoglobulin (IVIG). Meanwhile, pulmonary infections were more common, prevalence of hospitalized respiratory failure rate was higher in the poor LIPI group than that in the good LIPI group. After a median follow-up of 117 weeks, a total of 84 (10.1%) patients died in the first year and all-cause mortality was significantly higher in patients with poor LIPI (*P*<0.01).

Kaplan–Meier curves showed that 1-year survival rate were significantly higher in patients with good LIPI than those in the poor LIPI group (Figure 2). Multivariate Cox analysis (Table 2) showed that, compared to patients with poor LIPI, the adjusted *HR* for all-cause 1-year mortality was 0.22 (95% *CI* 0.05-0.93) for patients in the good LIPI group. After excluding patients who were lost for follow-up within one year, we found a similar result (Table 3, *HR* 0.20; 95% *CI* 0.05-0.86; *P*=0.03).

4. Discussion

In this study, we found a significant association between baseline LIPI and the risk of 1-year mortality in a large Chinese hospital-based cohort. The results indicated that, for patients diagnosed with IIM-ILD, good LIPI independently decreases the risk of all-cause mortality in 1-year by around 88% using poor LIPI as the reference. To the best of our knowledge, this is the first study exploring the association between LIPI and mortality in IIM-ILD cohort.

There were several explanations for the higher mortality rate in patients with poor LIPI. First, more recurrent patients were included in the poor LIPI group in this study. This result is consistent with a previous study conducted in China in which Chen and colleagues analyzed data of 132 patients with antisynthetase syndrome associated interstitial lung disease (ASS-ILD), the results showed that patients with recurrence had a higher mortality, and they also found serum LDH as an independent risk factor for the progression of ASS-ILD[14]. Besides, we found more patients had high CCI in poor LIPI group, and they had lower ALB, which could partially explain the higher mortality in this group.

At present, there is no published guideline for management of IIM-ILD, and patients are treated based on physicians' clinical experience and preference[1]. While there is a trend that high doses of corticoid, intravenous use of cyclophosphamide, IVIG, corticoid-plus would be used in patients in worse condition, like had more extra-muscle involvement and less clinical responses for first-line therapy[15]. This study showed that more patients received high doses of corticoid and IVIG in poor LIPI group, which indicates that they might have more activated muscle inflammation and worse pulmonary progression. Meanwhile, higher pulmonary infection rate and higher respiratory failure rate in the poor LIPI group could also support that.

Our results contrast with a previous study which reported that dermatomyositis associated interstitial lung disease (DM-ILD) and clinically amyopathic dermatomyositis associated interstitial lung disease (ADM-ILD) had significantly lower survival rates[16], while this study reported a higher proportion of ADM and ASS patients

with a lower mortality in the good LIPI group. This contrast could be explained as follows: Elevations of creatines, like serum creatine kinase and aspartate transaminase/alanine transaminase ratio, are common in IIM patients because it affects skeletal muscle, and their levels may partially indicate extent of muscle damages. While for extra-muscle predominance, more damages was observed in extra-muscle organs but there were less damage in skeletal muscle, which means creatines levels would be normal or slightly high. Previous studies have shown that IIM patients with extra-muscle manifestations had a higher mortality than those had no extra-muscle symptoms[17,18]. That would be the "creatine-mortality separation" for IIM patients, which means lower creatine and higher mortality. In this study, "LDH<250 IU/L and dNLR<3" was considered as the the criteria of good LIPI, and ADM and ASS were mainly characterized by extra-muscle manifestation, so it's reasonable that more ADM and ASS patients were included in the good LIPI group.

The strength of this study is the large sample size, while this study has some limitations: (1) the retrospective nature restricts a comprehensive data collection which would be helpful to describe the cohort better, for example, we do not have lung function test and pulmonary hypertension data to show how severe ILD is in our patients; (2) this study is based on West China hospital which is a first-class hospital in China, and most patients included in this study are severe and have received sevearl kinds of treatments, so the generalization of our results is restricted; (3) as in all single-center studies, the selective bias of patients enrollment in this study can not be avoided. Hence, more multi-center prospective studies are expected in the future to validate the results, and explore LIPI as a user-friendly biomarker for IIM-ILD patients in clinical works.

In conclusion, good LIPI was independently associated with decreased risk of all-cause 1-year mortality in IIM-ILD patients. This easy-to-obtain index might be served as a potential marker for assessing the prognosis of IIM-ILD.

Conflict of interest statement

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Authors' contributions

All authors contributed to the final manuscript. DL analyzed the data and drafted the manuscript. Corresponding authors Qiaoli Su read and revised the manuscript. ZZ, WZ performed the statistical analysis. CL, WZ, WZ, LH, HY collected the clinical data. All authors have read and approved the final submitted version.

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