Section: Miscellaneous

Original Research Article

HIGH-RESOLUTION CT EVALUATION OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS: INSIGHTS INTO CLASSIC AND VARIANT UIP PATTERNS FROM A SOUTH INDIAN TERTIARY CENTRE

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ABSTRACT

Background: Objective: To study the patterns of interstitial lung disease (ILD) in patients with systemic sclerosis (SSc) using high-resolution CT (HRCT), and to look at both classic and variant fibrotic features in patients with usual interstitial pneumonia (UIP) patterns in a South Indian tertiary care hospital. **Materials and Methods:** HRCT scans of 35 SSc patients collected over five years were analyzed. ILD patterns were classified as NSIP, UIP, or indeterminate. Variant fibrotic signs—straight-edge, four-corners, anterior upper lobe, and exuberant honeycombing—were recorded in UIP cases. Patients with "classic" UIP without variant signs were also noted. Two radiologists independently reviewed all scans. **Result:** ILD was seen in 34 out of 35 patients (97%). NSIP was the most common pattern (24/35, 69%), while UIP was present in 10 patients (29%). Among UIP patients, 4 (40%) showed classic UIP without variant signs, and 6 (60%) had 1–2 variant signs: straight-edge (3 patients), four-corners (2), anterior upper lobe (1), and exuberant honeycombing (2). No patient had all four variant signs. Inter-observer agreement was excellent (kappa = 0.83). **Conclusion:** ILD is very common in SSc patients in South India, with NSIP being the dominant pattern. UIP can appear with or without variant fibrotic signs. Recognizing both classic and variant UIP patterns on HRCT helps radiologists make accurate diagnoses and could guide prognosis.

INTRODUCTION

Systemic sclerosis (SSc) is an autoimmune disease that affects the skin and internal organs. The lungs are often involved, and interstitial lung disease (ILD) is the main cause of illness and death in these patients.[1-3] High-resolution CT (HRCT) is the best imaging tool to identify ILD and to tell apart different patterns like nonspecific interstitial pneumonia (NSIP) and usual interstitial pneumonia (UIP).[4,5]

NSIP is the most common ILD pattern in SSc and generally has a better prognosis. UIP, on the other hand, carries a higher risk and may progress more rapidly.[6,7] Variant fibrotic signs—such as straight-edge, four-corners, anterior upper lobe, and exuberant honeycombing—are sometimes seen in UIP related to SSc. These signs can help distinguish SSc-UIP from idiopathic UIP.[8-12]

However, not all UIP patients have these variant signs. Some patients show classic UIP features without any of the extra signs. Few studies from South India have prospectively looked at both classic and variant UIP in SSc patients. This study aims to fill that gap.

MATERIALS AND METHODS

Study Design

This was a **prospective study** conducted at Malabar medical college from 2019 to 2024. Thirty-five consecutive adult patients with SSc, meeting the 2013 ACR/EULAR criteria, were included. Patients with other lung diseases or poor-quality CT scans were excluded.

HRCT Protocol

All patients underwent volumetric HRCT scans with 1 mm slices. Scans were done during full inspiration, without contrast.

Image Review

Two experienced radiologists reviewed the scans independently. ILD patterns were classified as:

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- **NSIP:** Mainly ground-glass opacities, sometimes with subpleural sparing
- **UIP:** Fibrosis in basal and subpleural regions, reticulations, traction bronchiectasis, and honeycombing
- **Indeterminate/probable UIP:** Features not fully fitting UIP

Variant fibrotic signs were recorded in UIP cases:

- Straight-edge
- Four-corners
- Anterior upper lobe
- Exuberant honeycombing

Patients with UIP but without any of these variant signs were labelled as **classic UIP**. Disagreements were resolved by consensus.

RESULTS

Patient Characteristics

- Total patients: 35
- Females: 27 (77%)
- Males: 8 (23%)
- Mean age: 46 ± 10 years
- Median disease duration: 5.8 years

Table 1: HRCT Findings

Pattern	Number (%)
NSIP	24 (69%)
UIP	10 (29%)
Indeterminate	0

Table 2: UIP pattern

UIP Type	Number (%)	Variant Signs
Classic UIP (no variants)	4 (40%)	None
UIP with variants	6 (60%)	Straight-edge 3, Four-corners 2, Anterior upper lobe 1, Exuberant honeycombing 2

Inter-observer agreement: **kappa = 0.83** (excellent)

DISCUSSION

Interstitial lung disease is very common in SSc. In our study, 97% of patients had ILD on HRCT, which aligns with previous research.[1-3] NSIP was the most frequent pattern, as expected, while UIP appeared in almost one-third of patients.

A key observation was that some UIP patients did not show any variant fibrotic signs. Four patients had classic UIP, meaning their scans showed the typical basal and subpleural fibrosis, reticulation, and honeycombing—but none of the extra features like straight-edge or four-corners. This confirms that variant signs are helpful but not required to identify UIP.[8-10]

Among the six patients with variant signs, most had one or two, and no patient showed all four. This suggests that variant signs are complementary and may highlight subtle differences in disease distribution, but are not universally present.

Recognizing both classic UIP and variant UIP is important. Classic UIP without variant signs is still a serious fibrotic pattern and requires careful follow-up, just like UIP with variant signs. Radiologists should therefore focus on the core features of UIP first, and then note any additional variant signs.

Our study also shows excellent agreement between radiologists, confirming that HRCT is a reliable tool for identifying and classifying SSc-ILD.[11,13]

While NSIP remains the dominant pattern and generally responds better to immunosuppressive therapy, UIP—whether classic or with variant signs—often progresses faster and may require antifibrotic treatment.[6,7,12] Recognizing the spectrum of UIP appearances helps guide patient management and counseling.

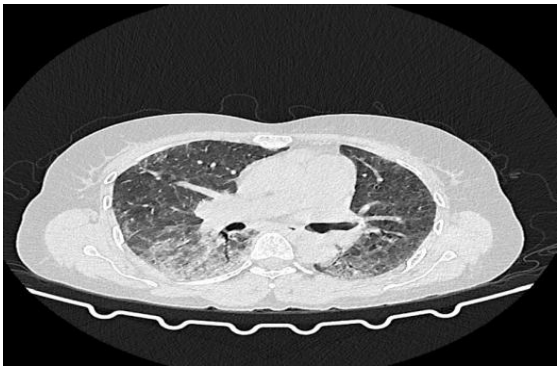

Figure 1: Case of scleroderma with NSIP pattern

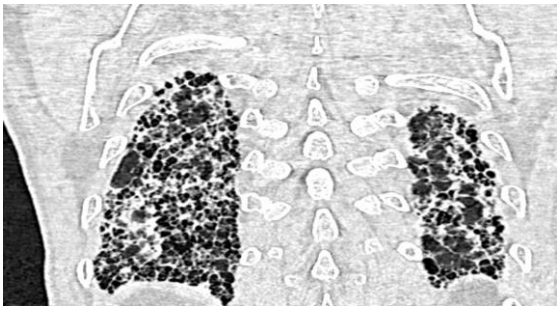

Figure 2: Case of exuberant honeycombing with variant fibrotic pattern

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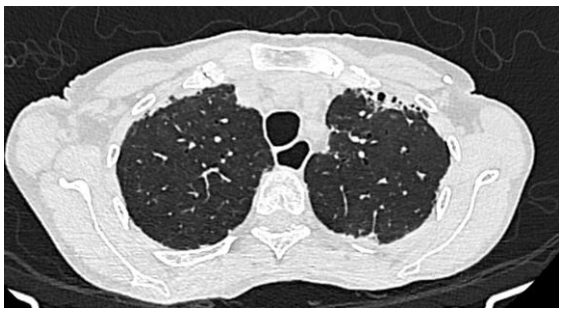

Figure 3: Case of anterior upper lobe sign

CONCLUSION

In South Indian patients with systemic sclerosis, ILD is highly prevalent. NSIP is the most common pattern, while UIP appears in about one-third. Among UIP patients, some show classic UIP without variant signs, while others display 1–2 variant fibrotic signs. Recognizing both classic and variant UIP on HRCT is crucial for accurate diagnosis and management.

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