The different faces of Nonspecific Interstitial Pneumonia

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Authors: M. Simões; Lisbon/PT
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Learning objectives

Idiopathic interstitial pneumonias consist in a group of lung diseases that are characterized by the presence of inflammation and fibrosis, that affect the lungs in varying degrees.

Nonspecific interstitial pneumonias (NSIP) is the second most common form of idiopathic pulmonary fibrosis, accounting for 14-36% of cases (1), being usual interstitial pneumonias (UIP) the most frequent.

Although interstitial lung diseases have similar clinical and radiologic correlations, it's most important to achieve a correct diagnosis, mainly due to different treatment and prognosis.

The role of high resolution computed tomography (HRCT) is well known from several studies, although the diagnosis of NSIP is frequently difficult mainly due to the overlap of radiologic findings seen in other interstitial lung diseases.

The goal of our work is to review radiologic findings on HRCT seen in nonspecific interstitial pneumonia (NSIP), as well as demonstration of lung abnormalities changes that occur over time in this entity.

In addition, less frequent findings of the disease are presented that may lead toward different diagnosis.
Background

As in most cases of lung interstitial diseases, clinical findings in NSIP are nonspecific, being the most prominent symptoms the gradual onset of dyspnea and cough. Most patients have between 40 to 50 years old, with no prevalence of gender. There is no known association with smoking.

Although many cases of NSIP are idiopathic, there is an association with several collagen vascular diseases namely: scleroderma, polyomyositis/dermatomyositis, Sjögren syndrome and rheumatoid arthritis as well as systemic lupus erythematosis. Not only it is important to look for associated findings that might be present on the CT scan, one has to be reminded that collagen diseases may precede the clinical presentation of the lung disease, sometimes by more than 5 years (2). In addition, NSIP can also be associated with drugs and occupational exposure (3).

NSIP is commonly characterized by temporally and spatially homogeneous lung involvement, being the key factor for the differentiation between NSIP and other interstitial lung diseases.

The two main histologic types of this disease are divided in cellular and fibrotic forms that differ from one another in the degree of inflammation and fibrosis.
Imaging findings OR Procedure details

Chest radiography is frequently normal or may show subtle changes in early stages of NSIP (Figure 1), although in advanced disease, there may be bilateral areas of infiltrates (Figure 2).

On HRCT, the most common abnormality consists in the presence of patchy areas of ground-glass opacities which have been reported in about 76-100% of cases (4) (Figures 3 and 4) (Figures 5, 6 and 7). The presence of ground glass opacities may also be a dominant feature of other lung diseases, but its absence helps to rule out NSIP and may lead towards alternative diagnosis (Figures 8 and 9) (Figures 10 and 11).

Irregular linear or intralobular reticular opacities combined with dispersed micronodules are findings most frequently seen in the fibrotic subtype of NSIP (Figure 12). There is typically a subpleural distribution of lung abnormalities, although many studies have shown rather a significant variability in its distribution (5).

Relative sparing of the subpleural region in the dorsal lung regions of the lower lobes is seen in up to 43% of patients (6) (Figure 13 and 14).

Symmetry of lung changes is another key factor that frequently can help the diagnosis, as the lung changes are fairly symmetric, with the lower lobes usually being more involved than the upper, without an apicobasal gradient seen as a rule in UIP (Fig 15 and 16).

In cases of advanced stages of disease or in fibrotic NSIP, traction bronchiectasis and reticulation may be a dominant feature, being most prominent in the lower lung zones (Figures 17 and 18). These findings may be seen in several other fibrotic lung diseases, as in hypersensitive pneumonitis or UIP, and is thus of limited diagnostic value (Figure 19) (Figures 20 and 21).

Focal areas of organizing pneumonia seen as areas of consolidation may appear in up to 50% of patients with NSIP, but is not a dominant finding (7) (Figure 22).

Other findings of NSIP include the presence of subpleural cysts, being usually of small size and of limited extended as compared to UIP (Figures 23 and 24).
The key HRCT features in NSIP are symmetry of lung changes with a basal predominance, characterized by the presence of ground glass opacities, mild reticulation, traction bronchiectasis and microcystic honeycombing with relative subpleural sparing.

Given the association with collagen vascular diseases, as previously stated, it is important to look for associated abnormalities on CT that may help in the diagnosis. Findings of an underlying collagen disease include the presence of an enlarged esophagus as seen in scleroderma, presence of pericardial or pleural effusion, seen in lupus or enlargement of the pulmonary arteries as a sign of pulmonary hypertension in patients with rheumatoid arthritis or scleroderma.
Images for this section:

Fig. 1: Chest radiograph: Moderate decrease in lung volumes in a patient with NSIP. No significant parenchymal abnormalities are seen.

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Fig. 2: Chest radiograph: Patient with NSIP showing decrease in lung volumes along with bilateral parenchymal infiltrates predominantly in the lower 2/3 of the lungs.

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Fig. 3: HRCT with characteristic findings of NSIP: Presence of ground glass opacities in the lower lobes with minimal subpleural reticulation.

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Fig. 4: HRCT with characteristic findings of NSIP: Presence of ground glass opacities in the lower lobes with minimal subpleural reticulation and traction bronchiectasis and bronchiolectasis.

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**Fig. 5:** HRCT in a patient with NSIP showing bilateral ground glass opacities with minimal subpleural reticulation. One month later (Fig 6) a marked increase in extent of ground glass opacities is seen affecting both lungs as a sign of acute exacerbation. These findings would significantly improve over time with treatment as it is seen on CT 12 months later(Fig 7).

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**Fig. 6:** HRCT in a patient with NSIP showing bilateral ground glass opacities with minimal subpleural reticulation. One month later (Fig 6) a marked increase in extent of ground glass opacities is seen affecting both lungs as a sign of acute exacerbation. These findings would significantly improve over time with treatment as it is seen on CT 12 months later (Fig 7).

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**Fig. 7:** HRCT in a patient with NSIP showing bilateral ground glass opacities with minimal subpleural reticulation. One month later (Fig 6) a marked increase in extent of ground glass opacities is seen affecting both lungs as a sign of acute exacerbation. These findings would significantly improve over time with treatment as it is seen on CT 12 months later (Fig 7).

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**Fig. 8:** Ground glass opacities in a patient with COP: Fig 8 shows in the lower left lobe an area of central ground-glass opacity surrounded by a denser area of consolidation known as the reversed halo sign or atoll sign (blue arrow). Areas of consolidation and ground glass are present in both lungs (Fig 9). Note the absence of reticulation and traction bronchiectasis that should point towards an alternative diagnosis.

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**Fig. 9:** Ground glass opacities in a patient with COP: Fig 8 shows in the lower left lobe an area of central ground-glass opacity surrounded by a denser area of consolidation known as the reversed halo sign or atoll sign (blue arrow). Areas of consolidation and ground glass are present in both lungs (Fig 9). Note the absence of reticulation and traction bronchiectasis that should point towards an alternative diagnosis.

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Fig. 10: Diffuse and ill defined centilobular nodules on both lungs in a patient with hypersensitivity pneumonitis. The presence of centrilobular nodules is an infrequent finding in NSIP and should suggest another diagnosis.

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**Fig. 11:** Difuse and ill defined centilobular nodules on both lungs in a patient with hypersensitivity pneumonitis. The presence of centrilobular nodules is an infrequent finding in NSIP and should suggest another diagnosis.

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Fig. 13: Area of focal consolidation in the upper right lobe in a patient with NSIP. Areas of consolidation are uncommon in patients with NSIP and cases of chronic consolidation are often associated to cryptogenic organizing pneumonia (COP).

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Fig. 14: Subpleural sparing in NSIP seen in a patient with scleroderma. HRCT shows lower lobe ground glass opacities with mild reticulation with relative sparing of the subpleural lung (blue arrows). This finding reinforces the diagnosis of NSIP, although its absence doesn’t exclude it. Also note the enlarged esophagus in figure b (orange arrow).

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Fig. 15: Subpleural sparing in NSIP seen in a patient with scleroderma. HRCT shows lower lobe ground glass opacities with mild reticulation with relative sparing of the subpleural lung (blue arrows). This finding reinforces the diagnosis of NSIP, although its absence doesn’t exclude it. Also note the enlarged esophagus in figure b (orange arrow).

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**Fig. 16:** Patient with NSIP: On coronal view one can see lower lobe predominance of the changes. Small cystic lesions can also be depicted.

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Fig. 17: Patient with NSIP: Diffuse lung involvement with irregular linear reticulation predominantly at the periphery of the lungs and basal ground glass opacities. Note the cystic lesions in the subpleural region.

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Fig. 18: Fibrotic NSIP showing lower lobe predominant lung changes with reticulation, ground glass opacities and traction bronchiectasis (orange arrows).

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Fig. 19: Fibrotic NSIP showing lower lobe predominant lung changes with reticulation, ground glass opacities and traction bronchiectasis (orange arrows).

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**Fig. 20:** Patient with chronic hypersensitivity pneumonitis. Reticulation and honeycombing are predominant features (black arrows). Note the absence of areas of ground glass in this case as compared with the cases of NSIP.

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**Fig. 21:** Patient with UIP: HRCT shows predominant honeycombing in the lower lobes without any ground glass opacities. The extent of honeycombing in this case along with the absence of ground glass, is not a characteristic of NSIP and should point towards different diagnosis.

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**Fig. 22:** Patient with UIP: HRCT shows predominant honeycombing in the lower lobes without any ground glass opacities. The extent of honeycombing in this case along with the absence of ground glass, is not a characteristic of NSIP and should point towards different diagnosis.

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**Fig. 23:** Patient with fibrotic NSIP: Lower lobe reticulation of the lung parenchyma along with areas of ground glass opacity, traction bronchiectasis and microcystic changes. The left lower lobe has more predominant changes (better depicted in figure 24).

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Fig. 24: Patient with fibrotic NSIP: Lower lobe reticulation of the lung parenchyma along with areas of ground glass opacity, traction bronchiectasis and microcystic changes. The left lower lobe predominance.

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Fig. 12: Patient with fibrotic NSIP showing focal areas of ground glass and a few scattered micronodules (orange arrows). Micronodularity is not a prominent feature in NSIP.

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Conclusion

In the majority of patients with NSIP, lung abnormalities are characterized by ground glass opacities along with relatively mild reticulation of the lower lobes with sparing of the subpleural space.

No single radiologic finding is diagnostic of NSIP, but the constellation of these findings in the appropriate clinical setting can help in the diagnosis. Howbeit, the best approach for interstitial pneumonias lies in a general consensus between clinicians, radiologists and pathologists.
References


