CASE REPORT

Cellular non-specific interstitial pneumonia masquerading as congestive heart failure

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Dr Takeshi Saraya,
sara@yd5.so-net.ne.jp**SUMMARY**

A 66-year-old woman with a history of myocardial infarction 2 months prior presented to our respiratory department with several days of dry cough and night sweats. Chest X-ray and thoracic CT showed ground glass opacities or consolidation spreading from the hilar area to the peripheral area, suggesting central redistribution. Although neither rales nor abnormal heart sounds were noted, she was tentatively diagnosed with congestive heart failure based on those radiological findings. However, radiographic lung lesions and her symptoms were refractory to intensive diuretic treatment. Thereafter, video-assisted thoracoscopic surgery was performed, resulting in a diagnosis of cellular non-specific interstitial pneumonia (c-NSIP). After initiating treatment with prednisolone, her symptoms and the radiological findings resolved. In patients with NSIP, a radiological central distribution could rarely occur, especially in cases of c-NSIP. No rales were detected because of its paucity of fibrous components in the lung.

BACKGROUND

Idiopathic non-specific interstitial pneumonia (NSIP) is a rare clinical disorder and it can be classified into cellular NSIP (c-NSIP) or fibrotic NSIP patterns. Both clinical conditions manifest with ground glass opacities (GGO), irregular linear opacities, and consolidations in a bilateral, symmetrical or subpleural distribution. The present report described a rare case of c-NSIP that presented with

lung involvement in a central distribution, thereby radiologically mimicking congestive heart failure.

CASE

A 66-year-old woman was referred to our respiratory department with several days of dry cough and night sweats. She had been treated for essential hypertension, hyperlipidaemia and atrial fibrillation since 7 years. Her regular medications were aspirin (100 mg/day), bisoprolol fumarate (2.5 mg/day), candesartan cilexetil (4 mg/day), atorvastatin calcium hydrate (10 mg/day) and spironolactone (25 mg/day) and her regimen had remained stable over the 7-year treatment period. She experienced myocardial infarction 2 months prior to the current presentation and was successfully treated with non-drug eluting stent insertion for left anterior descending artery stenosis. She was a housewife and an ex-smoker with a history of 30 pack-years. She denied any dust exposure or sick contacts, and no paroxysmal nocturnal dyspnoea was noted. Her vital signs were as follows: blood pressure 160/80 mm Hg, heart rate of 58 bpm, respiratory rate of 18 breaths/min, body temperature of 36°C and oxygen saturation of 97% measured at ambient air. Physical examination showed bilateral earlobe creases, point of maximal impulse that was shifted 2.5 cm to the left side from left mid-clavicular line at the left fifth-intercostal space. However, jugular venous pressure was normal (8 cm) and neither jugular venous dilatation nor hepatojugular reflux was noted. No rales or abnormal heart sounds

Figure 1 Chest X-ray taken 3 months earlier (A) appeared to be normal (CTR, cardiothoracic ratio, 54.2%). At first visit to our respiratory department, chest X-ray (B) showed bilateral infiltrates spreading from the hilar area to the peripheral area with mild cardiomegaly (CTR 59.2%).

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Figure 2 Non-enhanced thoracic CT (A and B) depicted bilateral ground glass opacities and consolidation expanding from the hilar region, compatible with congestive heart failure.

were recognised. Bilateral slight pitting oedema was noted below each knee. Chest X-ray at our department (figure 1B) showed bilateral infiltrates spreading from the hilar area to the peripheral area with mild cardiomegaly (CTR, cardiothoracic ratio, 59.2%), which seemed to be larger than that assessed 3 months earlier (figure 1A; CTR 54.2%). Although no typical X-ray signs (eg, Kerley A, B or C lines) for congestive heart failure¹ were noted, she was admitted to our cardiology ward with a tentative diagnosis of congestive heart failure. Non-enhanced thoracic CT (figure 2A, B) depicted bilateral GGO and consolidation expanding from the hilar region, compatible with congestive heart failure. Serum laboratory data on admission was normal with the exception of mild elevation of white cell count (10 500/ μ L), blood urea nitrogen (28 mg/dL), B-type natriuretic peptide (78 pg/mL) and creatinine (1.2 mg/dL), as well as a marked elevation of Krebs von den Lungen-6 (KL-6, 1539 IU/L).

After admission, intensive diuretic therapy was initiated, but echocardiography at 3rd hospital day showed normal ejection fraction of 57% and abnormal lung shadows persisted on re-evaluation performed on the 14th hospital day.

INVESTIGATIONS

No rales were noted throughout in the lung fields, but persistence of the lung lesions raised the possibility of idiopathic interstitial pneumonia. Therefore, further diagnostic evaluation was performed using video-assisted thoracoscopic surgery (VATS). Panoramic view (figure 3A) of biopsied specimens obtained

Figure 3 Panoramic view (A) of biopsied specimens obtained from right S3 showing the lung lesion located in the central area of the resected lung section. On H&E with high-power field (B) demonstrating a lymphocytic infiltration in the alveolar septa. A fibrous component on silver stain (C) was scarcely recognised.

from right S3 showed that the lung lesion was located in the central area of the resected lung section. Those lesions were stained with H&E and examined on high-power field (figure 3B), demonstrating a lymphocytic infiltration in the alveolar septa. A fibrous component on silver stain (figure 3C) was recognised, although it was faint. Those results were compatible with a diagnosis of c-NSIP.

TREATMENT

Prednisolone therapy (0.8 mg/kg/day) was initiated and the radiographic lung lesions completely disappeared within 1 month. She was discharged from the hospital uneventfully.

OUTCOME AND FOLLOW-UP

The treatment dose was tapered over 6 months in an outpatient setting and 2 years after withdrawal of treatment, she was still in good health with no recurrence of lung disease.

DISCUSSION

There are two primary forms of NSIP: cellular and fibrotic. The cellular form is defined mainly by inflammation of the cells of the interstitium, while the fibrotic form is defined by thickening and fibrosis of lung tissue. Although CT findings in cases of NSIP can vary, GGO or irregular linear opacities, peribronchovascular predominant fibrosis and consolidations with relative sparing of the immediate subpleural lung are typically seen in a bilateral and symmetrical distribution² as in the present case. However, GGO or consolidations are generally distributed in peripheral or subpleural spaces and not in the central area,² as well as lower lobe predominance.³ In this regard, the present case had an atypical radiological pattern for NSIP and the absence of late inspiratory crackles as well as a recent history of myocardial infarction led to an erroneous diagnosis of congestive heart failure. The absence of late inspiratory crackles was likely related to the paucity of fibrous change in the lung parenchyma, which was confirmed by the pathological assessment on VATS-biopsied specimens. Thus, at a glance, differentiation between heart failure and NSIP is difficult and needs multidisciplinary analysis on physical, radiological and clinical features. A CT taken at later phase (figures not shown) revealed

perivascular-prominent fibrosis and moderate traction bronchiectasis, which might be a clue to the diagnosis for NSIP rather than heart failure. This case suggests that NSIP cases can present with an atypical central distribution on radiological assessment. Further, in a patient with c-NSIP, the lack of late inspiratory crackles due to paucity of fibrous component in the lung parenchyma might be a clinical clue to the diagnosis.

Learning points

- ▶ In general, areas of glass opacities or consolidation in patients with non-specific interstitial pneumonia (NSIP) are distributed in bilateral or symmetrical fashion on radiological studies and are predominantly seen in the peripheral or subpleural spaces.
- ▶ A central distribution of lung involvement is rarely seen, but should be considered as an atypical radiological finding in patients with NSIP.
- ▶ In patients with cellular-NSIP, paucity of fibrous components in the lung parenchyma might result in normal auscultatory sounds, as in the present case.

Contributors TS, ST and MF wrote and discussed the case regarding the correlation between radiological and pathological findings. HT performed a surgery and managed the patient.

Competing interests None.

Patient consent Obtained.

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