|  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
|

|  |  |  |  |
| --- | --- | --- | --- |
| **Characteristics** | **CPFE with CTD****N=47** | **CPFE without CTD** **N=181** | **P value**\* |
| Number of male patients (%) | 32 (68.1) | 173 (95.6)  | <0.001 |
| Median age-years (IQR) | 68.0 (60.0, 70.5) | 71.0 (66.0, 77.0) | <0.001 |
| Former/Current smoker (%) | 26 (55.3)/21 (44.7) | 129 (71.3)/52 (28.7) | 0.080 |
| Pack-years smoking, median (IQR) | 45 (28, 68.8) | 51 (40, 80) | 0.035 |
| Serum KL-6, median U/mL (IQR) | 534 (308, 844) | 564 (383, 851) | 0.430 |
| Pulmonary hypertension (%) | 7 (14.9) | 19 (10.5)  | 0.440 |
| Lung cancer (%) | 3 (6.4) | 50 (27.6)  | 0.002 |
| Connective tissue disease |  |  |  |
| Rheumatoid arthritis (%) | 28 (59.6) |  |  |
| Systemic sclerosis (%) | 6 (12.8) |  |  |
| ANCA-associated vasculitis (%) | 4 (8.5) |  |  |
| Dermatomyositis (%) | 4 (8.5) |  |  |
| Systemic Lupus Erythematosus (%) | 4 (8.5) |  |  |
| Sjogren’s syndrome (%) | 1 (2.1) |  |  |
| Serological autoimmunity (%)**†** | 36 (76.6) | 45 (24.5) |  |
| Rheumatoid factor (%) | 25 (53.1) | 20 (11.0) |  |
| Anti-citrullinated protein antibody (%) | 15 (31.9) | 15 (8.3) |  |
| Anti-nuclear antibody (%) | 14 (29.8) | 20 (11.0) |  |
| Initial CT findings‡ |  |  |  |
| Total lung volume, median cm3 (IQR) | 4208.1 (3967.5, 5062.7) | 4418.3 (3787.4, 5250.3) | 0.919 |
| Extent of normal lung, median cm3 (IQR) %j | 3035.6 (2804.0, 3724.4), 76.4  | 3191.6 (2335.4, 3824.9), 76.3 | 0.953 |
| Extent of emphysema, median cm3 (IQR) %  | 693.1 (251.4, 1361.3), 17.0 | 574.6 (265.1, 1194.0), 13.8 | 0.437 |
| Extent of fibrosis, median cm3 (IQR) % | 173.1 (81.7, 532.4), 4.2 | 301.2 (143.5, 523.3), 6.8 | 0.089 |
| Extent of fibrosis |  |  | 0.161 |
| Fibrosis <5% (%) | 24 (51.1) | 65 (35.9) |  |
| Fibrosis 5–10% (%) | 8 (17.0) | 46 (25.4) |  |
| Fibrosis ≥10% (%) | 15 (31.9) | 70 (38.7) |  |

 |

*CPFE* combined pulmonary fibrosis and emphysema, *CTD* connective tissue disease, *IQR* interquartile range, *CT* computed tomography.

\* P values are reported for the differences between the fibrosis proportion groups, using a chi-squared test, Fisher exact test, t test, or Wilcoxon rank-sum test as appropriate.

**†** Each positivity was defined as follows: rheumatoid factor ≥30 U/ml, anti-citrullinated protein antibody≥4.5 EU/mL, ANA≥1:320 titer.

‡Calculated results from CALLIPER.