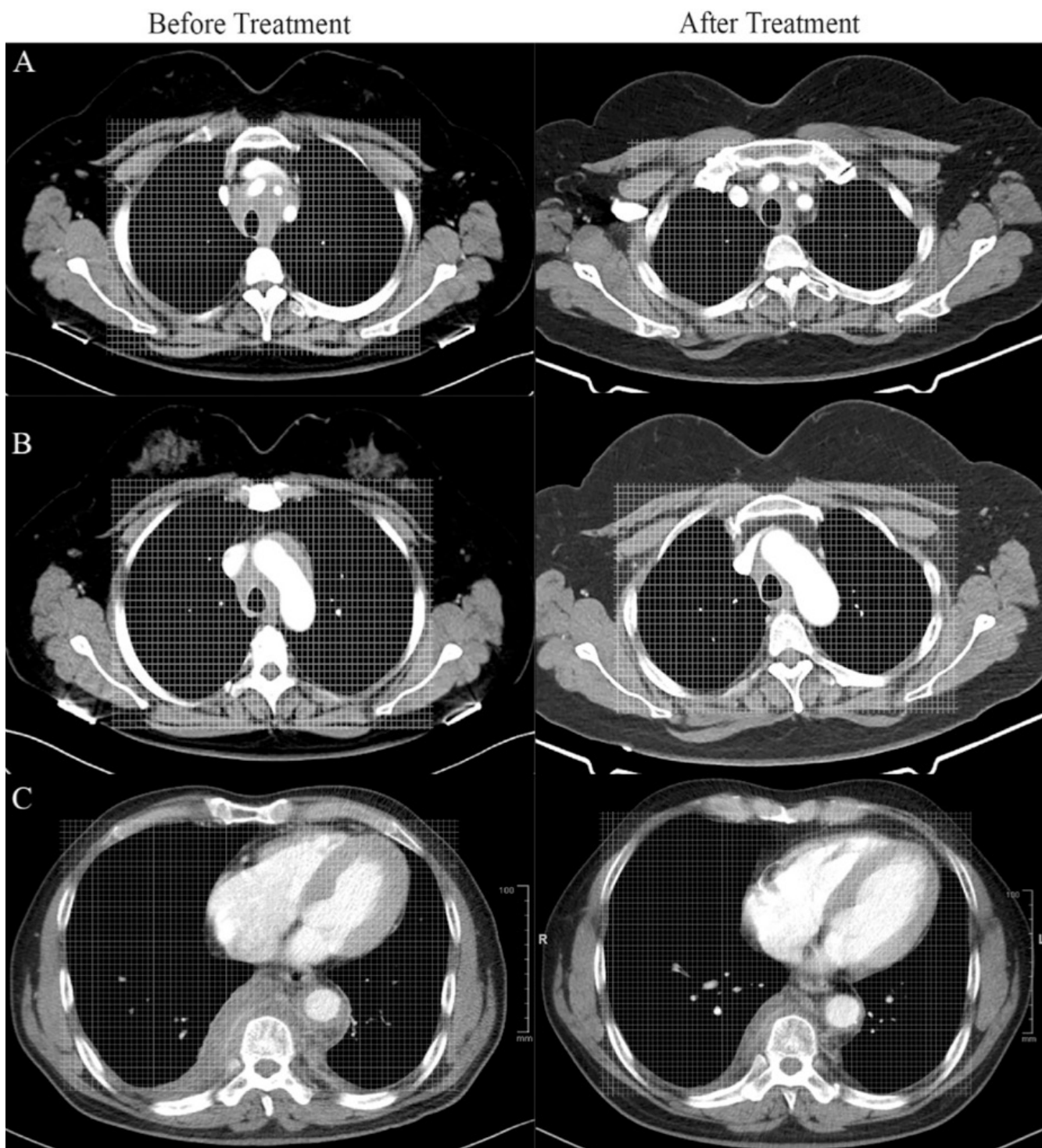


Supplementary file

Supplementary Table S1. Comparison of characteristics at baseline between the only FM (Group A) and FM patients with multiorgan involvement group (Group B).

Characteristics at baseline	Group A (n=7)	Group B (n=13)	<i>p</i> -value
Demographic features			
Age (years)	48.6 ± 16.1	51.2 ± 15.7	0.722
Male (n, %)	5 (71.4)	7 (53.8)	0.642
Disease duration (month), M (Q ₁ -Q ₃)	7 (1-12)	8 (0-11)	0.953
History of allergy (n, %)	0 (0)	3 (23.1)	0.521
Symptoms at disease onset (n, %)			
Chest pain	3 (42.9)	1 (7.7)	0.101
Back pain	2 (28.6)	3 (23.1)	1.000
Hoarseness of voice	2 (28.6)	2 (15.4)	0.587
Cough	0 (0)	4 (30.8)	0.249
Lymph node swelling	0 (0)	4 (30.8)	0.249
Submandibular gland enlargement	0 (0)	3 (23.1)	0.521
Arthralgia	0 (0)	3 (23.1)	0.521
Lacrimal gland enlargement	0 (0)	2 (15.4)	0.521
Lower limb oedema	0 (0)	2 (15.4)	0.521
Fever	0 (0)	2 (15.4)	0.521
Dysphagia	0 (0)	1 (7.7)	1.000
Laboratory parameters			
HGB (g/L)	137 ± 27	132 ± 17	0.383
WBC (10 ⁹ /L)	8.13 ± 3.18	7.71 ± 2.72	0.181
PLT (10 ⁹ /L)	271 ± 115	281 ± 50	0.262
ESR (mm/h), M (Q ₁ -Q ₃)	44 (8-87)	48 (31-65)	0.692
hsCRP (mg/L), M (Q ₁ -Q ₃)	31.14 (2.32-55.93)	6.75 (2.26-10.00)	0.181
IgG (g/L)	21.29 ± 6.96	22.84 ± 7.98	0.024*
IgA (g/L)	3.25 ± 0.87	2.77 ± 1.07	0.011*
IgM (g/L)	1.25 ± 0.67	1.33 ± 1.04	0.821
IgG1 (mg/L), M (Q ₁ -Q ₃)	11903 (8020-13100)	11246 (9810-13275)	0.308
IgG2 (mg/L), M (Q ₁ -Q ₃)	6679 (3920-8500)	6238 (3663-8575)	0.640
IgG3 (mg/L), M (Q ₁ -Q ₃)	586 (191-743)	512 (218-809)	0.350
IgG4 (mg/L), M (Q ₁ -Q ₃)	2799 (1440-4770)	10901 (989-23400)	0.303
T-IgE (KU/L), M (Q ₁ -Q ₃)	262.3 (17-655)	457.6 (71.0-520.0)	0.043*
C3 (g/L)	1.099 ± 0.268	0.719 ± 0.331	0.006*
C4 (g/L)	0.183 ± 0.060	0.127 ± 0.098	0.230
Cr (μmol/L)	94 ± 33	85 ± 48	0.900
Disease activity			
Baseline IgG4-RD RI	4.3 ± 1.6	11.8 ± 5.8	0.004*
Baseline PGA	3.1 ± 0.4	6.8 ± 2.6	0.002*

Q1: quartile 1; Q3, quartile 3; SD: standard deviation; WBC: white blood cell; HgB: haemoglobin; PLT: platelet; EOS: eosinophils; ESR: estimated sedimentation rate; hsCRP: hypersensitive C-reactive protein; Ig: immunoglobulin; C3: complement 3; C4: complement 4; Cr: creatinine; IgG4-RD RI: immunoglobulin G4-related disease responder index; T-Ig: total immunoglobulin.



Supplementary Fig. S1. Methods in treatment efficacy evaluation of IgG4-RFM patients.

Aortic root, aortic arch, descending aorta and thoracic vertebral were selected as three standard slices for treatment efficacy evaluation. After treatment, the mediastinal soft tissue at the level of aortic root, aortic arch, descending aorta and thoracic vertebral was decreased by 58% (A), 55% (B) and 59% (C), respectively. A and B was from patient 17, so the reduction was calculated as 56.5%. The reduction of soft tissues in patient 12 was recognised as 59% in C. Treatment efficacy of patient 17 and 12 was classified into remission of soft tissues by 31-70%.