

1 **ADDITIONAL FILE**

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3 **SUPPLEMENTARY TABLES**

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5 **Table S1: Comparison of the history of pulmonary involvement between Sjo-AD and**
6 **controls**

	Sjo-AD, n=31 (%)	Sjo controls, n=62 (%)	P
Articular involvement	18 (58.1)	34 (54.8)	0.83
Purpura	8 (25.8)	7 (11.3)	0.13
Muscular	3 (9.7)	3 (4.8)	0.40
Peripheral neuropathy	3 (9.7)	12 (19.3)	0.37
Central nervous system	1 (3.2)	5 (8.1)	0.66
Renal impairment	3 (9.7)	3 (4.8)	0.40
Splenomegaly	2 (6.4)	2 (3.2)	0.60
Adenopathy	7 (22.6)	6 (9.7)	0.12

7 Mann-Whitney test was used. $p < 0.05$ was considered significant.

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13 **Table S2: Description of Sjo-AD biological domain of ESSDAI**

	Sjo-AD, n=31 (%)
<u>Biological activity</u>	
-Absent	10 (32.3)
-Low	6 (19.3)
-Moderate	13 (41.9)
<u>Items</u>	
-Hypocomplementemia	7/24 (29.2)
-IgG level >16 and <20 g/L	3/25 (12)
-IgG level >20 g/L	8/25 (32)
-Hypergammaglobulinemia > 13.5 g/L	18/27 (66.7)
-Hypogammaglobulinemia < 8g/L	2/27 (7.4)
-Cryoglobulinemia	6/26 (23.1)

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17 **Table S3: Patients reported outcomes of Sjo-AD and controls**

	Sjo-AD, n=19 (%)	Sjo controls, n=57 (%)	p
VAS overall dryness	59.3 ± 19.8 [15]	51 ± 25	0.32
VAS asthenia	62.7 ± 28.8	63.2 ± 28.2 [56]	0.92
VAS pain	47.5 ± 30	53.7 ± 26	0.53

18 Data are presented as mean ± SD. The number of patients evaluated is indicated in square
19 brackets when data were missing. Mann-Whitney and Fisher's exact test were used. p<0.05
20 was considered significant.

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25 **Table S4: Sjo-AD evolution based on the CT pattern**

	Diffuse Sjo-AD, n=15 (%)	Localized Sjo- AD, n=8 (%)	p
Baseline PFT			
-FEV1 (%pred)	72.8 ± 18.8 [13]	93.9 ± 20.4	0.02
-FVC (%pred)	89.2 ± 19.5 [13]	106 ± 20.9	0.03
-FEV1/FVC	68.4 ± 9.9 [12]	74 ± 9.8	0.31
-TLC (%pred)	99.6 ± 14.8 [14]	94.3 ± 21.8	0.63
-DLCO (%pred)	68.1 ± 13.4 [12]	72.5 ± 13.3 [7]	0.43
Change in PFT (delta)			
-FEV1	-3.8 ± 15.5 [12]	-5.1 ± 10.2 [6]	0.68
-FVC	1.2 ± 7 [12]	-10.4 ± 12.2 [6]	0.05
-FEV1/FVC	-1.8 ± 4.9 [11]	2.6 ± 7.6 [6]	0.35
-TLC	-2.5 ± 11.8 [9]	4 ± 10.2 [6]	0.46
-DLCO	-10 ± 17.2 [7]	3.6 ± 9.4 [5]	0.09
CT scan evolution			
-Worsening	4/14 (28.6)	3/6 (50)	0.6
-Improvement/Stability	2/8/14 (71.4)	2/1/6 (50)	0.10

26 Diffuse Sjo-AD are defined as the association of both proximal (bronchiectasis) and distal
27 (bronchiolitis) airway disease while localized Sjo-AD is defined as proximal or distal airways
28 involvement. Delta represents the difference between last follow-up and baseline PFT. Mann-
29 Whitney and Fisher's exact test were used. The number of patients evaluated is indicated in
30 square brackets when data were missing. p<0.05 was considered significant.

31 **Table S5: Sjo-AD progression based on disease activity**

	ESSDAI>5, n=13 (%)	ESSDAI<5, n= 18 (%)	p
Baseline PFT			
-FEV1 (%pred)	71.5 ± 16.8 [12]	91.2 ± 22.4 [16]	0.02
-FVC (%pred)	87.9 ± 15.9 [12]	101 ± 23.6 [14]	0.08
-FEV1/FVC	68 ± 9.2 [11]	74.7 ± 10.5 [14]	0.12
-TLC (%pred)	95.2 ± 19.1 [12]	102.2 ± 21.2 [16]	0.73
-DLCO (%pred)	72.1 ± 14 [11]	69.4 ± 12 [14]	0.88
Change in PFT (delta)			
-FEV1	-1.1 ± 12.2 [10]	-3.8 ± 16.4 [10]	0.97
-FVC	-0.6 ± 12.3 [11]	0 ± 13 [9]	0.71
-FEV1/FVC	-0.7 ± 7.1 [10]	-0.9 ± 5.3 [9]	0.84
-TLC	3.2 ± 13.7 [9]	0.2 ± 9.7 [8]	0.42
-DLCO	-9.2 ± 21.9 [5]	-0.9 ± 9.5 [7]	0.85
CT scan evolution			
-Worsening	5/10 (50)	4/12 (33.3)	0.66
-Improvement/Stability	3/2/10 (50)	1/7/12 (66.7)	0.67

32 Delta represents difference between last follow-up and baseline PFT. Mann-Whitney and
 33 Fisher's exact test were used. The number of patients evaluated is indicated in square brackets
 34 when data were missing. p<0.05 was considered significant.

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47 **Table S6: Comparison between extensive and non-extensive Sjo-AD**

	Extensive Sjo-AD, n=9 (%)	Non-extensive Sjo-AD, n=13 (%)	p
Baseline PFT			
-FVC (%pred)	98.7 ± 22.5	93 ± 23.4 [11]	0.50
-FEV1 (%pred)	84.6 ± 16.6	77.1 ± 26.5 [11]	0.60
-FEV1/FVC	72.8 ± 8.3	68.6 ± 11.5 [10]	0.72
-TLC (%pred)	94.9 ± 22.1	99.4 ± 17.1 [12]	0.92
-DLCO (%pred)	76.3 ± 13.8 [8]	64.2 ± 10 [12]	0.02
Change in PFT (delta)			
-FVC	-8.3 ± 7.5 [6]	0.8 ± 11 [11]	0.01
-FEV1	-7.2 ± 8.9 [6]	-2.2 ± 16.3 [11]	0.22
-TLC	2.6 ± 13.4 [6]	-0.8 ± 10.7 [8]	0.66
-DLCO	-5.5 ± 11.7 [3]	-3.9 ± 17.3 [9]	0.68

48 Results are shown as n (%) unless otherwise indicated. Delta corresponds to the difference
 49 between last follow-up PFT and baseline PFT. Wilcoxon test was used. The number of
 50 patients evaluated is indicated in square brackets when data were missing. p<0.05 was
 51 considered significant.

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57 **Table S7: CT scan evolution based on ESSDAI and IgA status**

	ESSDAI>5 IgA+, n= 5 (%)	Negative score, n= 11 (%)	p
Worsening	5 (100)	4 (36.4)	0.03
Improvement/Stability	0/0	3/5 (72.7)	0.03

58 Fisher's exact test was used. p<0.05 was considered significant.

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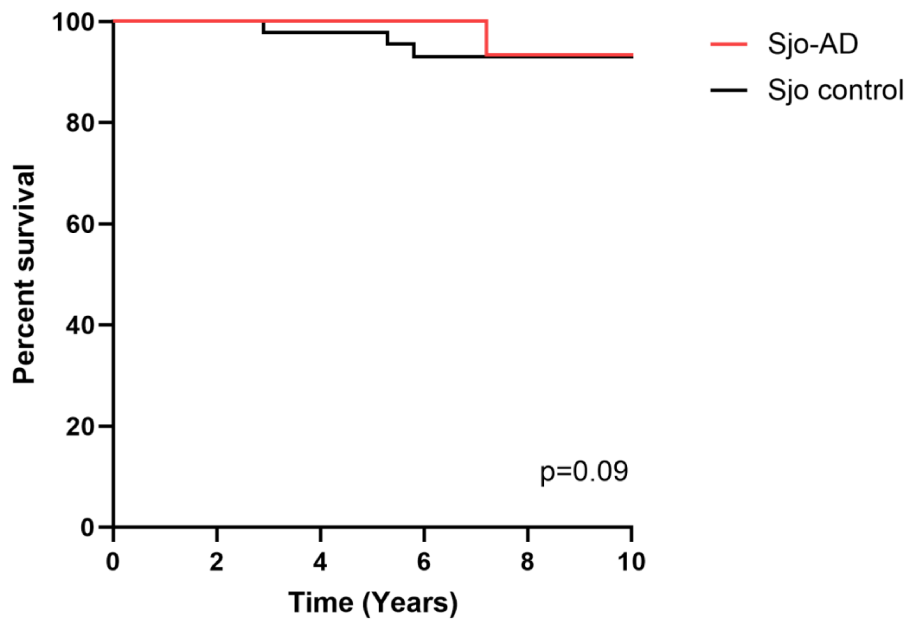
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67 SUPPLEMENTARY FIGURES

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**Number at risk**

Sjo-AD	27	27	22	17	12	10
Sjo control	49	46	44	39	27	6

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70 **Figure S1: Sjo-AD survival**

71 Kaplan Meier curves displaying the estimated survival probably was performed comparing
 72 Sjo-AD to control. The log-rank test was performed and indicates the difference between
 73 survival curves. $p < 0.05$ was considered significant.

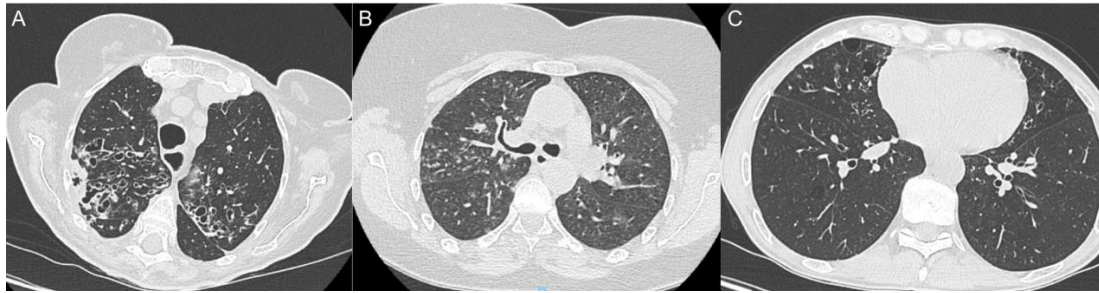
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80 **Figure S2: Chest CT scans typical aspects of Sjo-AD and cysts**

81 Illustrations of chest CT scan typical aspects of Sjo-AD and cysts. (A) Bronchiectasis (B)

82 Bronchiolitis (C) Cysts.