Radiological and clinical specific features of anti-synthetase syndrome : a monocentric retrospective analytic study

Q. MALOIR[1, 2] ; F. GESTER[1, 2] ; JL. CORHAY[1] ; R. LOUIS[1] ; J. GUIOT[1, 2]

(1) Department of respiratory medicine, CHU Liege, Belgium.

(2) Division of pulmonary interstitial and vascular diseases.

BACKGROUND

Anti-synthetase syndrome (ASS) is a rare auto-immune disorder combining autoantibodies and specifics clinical manifestations. One of the particularities of the ASS is the pleiomorphic radiological presentation seen at the initial work-up. Evaluating treatment response can also be challenging and requires a specific clinical, functional, biological and radiological monitoring.

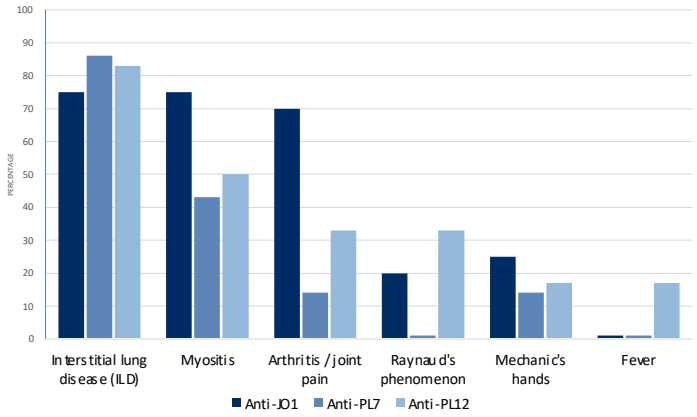
Aim of this study : To identify specific radiological and clinical features of ASS

PATIENTS CHARACTERISTICS

	All patients (n=33)	Anti-JO1 (n=20)	Anti-PL7 (n=7)	Anti-PL12 (n=6)	*p-value*
General characteristics:					
Male/Female	18 (55) / 15 (45)	9 (40) / 13 (60)	7 (100) / 0 (0)	3 (50) / 3 (50)	***0.021***
Age (years)	55.0 [range: 21-87]	53.1 [range: 26-87]	73.3 [range: 65-85]	39.8 [range: 21-62]	***0.0095***
Smokers	13 (39)	5 (30)	4 (57)	3 (50)	*0.34*
Proven deaths	11 (33)	6 (30)	3 (43)	2 (33)	*0.99*

Except where indicated, data are expressed as "number (%)"

Clinical characteristics :



References :

1. Lega JC, et al. — Idiopathic inflammatory myopathies and the lung. *Eur Respir Rev*, 2015, **24**, 216–238.
2. Fischer A, et al.— An official European Respiratory Society/American Thoracic Society research statement: Interstitial pneumonia with autoimmune features. *Eur Respir J,* 2015, **46**, 976–987.

METHOD

- We included 33 patients divided in three groups according to their autoimmune pattern (JO1, PL7 and PL12).
- We retrospectively studied all patients suffering from ASS in CHU of Liège until 2019. The diagnosis of ASS was made according to ATS/ERS recommandations[1,2]. We analyzed clinical features, pulmonary function tests (PFT), computed tomography (CT), and longitudinal evolution with regard to their treatments.

MAIN RESULTS

Initial HRCT findings

	All patients (n = 21)	Anti-JO1 (n = 13)	Anti-PL12 (n = 4)	Anti-PL7 (n = 4)
Ground glass opacities				
Grade A (Stades 0 - 1)	12 (57)	8 (62)	2 (50)	2 (50)
Grade B (Stades 2 - 4)	9 (43)	5 (38)	2 (50)	2 (50)
Reticulations				
Grade A (Stades 0 - 1)	10 (47)	7 (54)	2 (50)	1 (25)
Grade B (Stades 2 - 4)	11 (53)	6 (46)	2 (50)	3 (75)
Traction bronchectasis				
Grade A (Stades 0 - 1)	13 (62)	8 (62)	4 (100)	1 (25)
Grade B (Stades 2 - 4)	8 (38)	5 (38)	0 (0)	3 (75)
Consolidations				
Grade A (Stades 0 - 1)	16 (76)	9 (69)	4 (100)	3 (75)
Grade B (Stades 2 - 4)	5 (24)	4 (31)	0 (0)	1 (25)
Honeycombing				
Grade A (Stades 0 - 1)	19 (90)	12 (92)	4 (100)	3 (75)
Grade B (Stades 2 - 4)	2 (10)	1 (8)	0 (0)	1 (25)
Nodules	5 (24)			
Cysts	5 (24)			
Pleural effusion	3 (14)			

Data are expressed as "number (%)"

HRCT longitudinal analyses

Change in lesions between initial and follow-up CT (n = 11)	Increase or appear	No change	Decrease	Resolved	Initial CT evaluation (grade A / B)	Follow up CT evaluation (grade A / B)
GGO	0 (0)	5 (45)	5 (45)	1 (9)	6 / 5	9 / 2
Reticulations	0 (0)	10 (90)	1 (9)	0 (0)	5 / 6	6 / 5
Traction bronchiectasis	2 (18)	8 (73)	1 (9)	0 (0)	8 / 3	8 / 3
Consolidations	0 (0)	7 (64)	2 (18)	2 (18)	6 / 5	10 / 1
Honeycombing	1 (10)	10 (90)	0 (0)	0 (0)	11 / 0	10 / 1

Data are expressed as "number (%)"

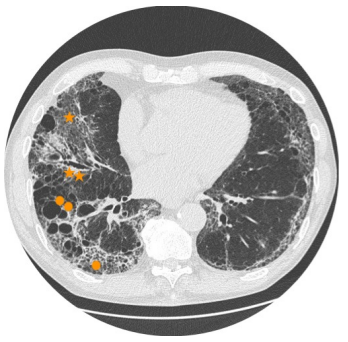


Figure 1 : Initial HRCT from patient with anti-PL12
★: Ground glass opacities; ★★: Traction bronchectasis; ●: Honeycombing; ●●: Cysts; ■: Subpleural reticulations; ■■: Pleural effusion

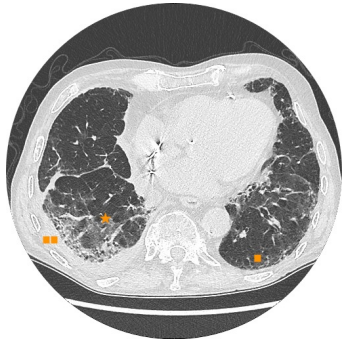


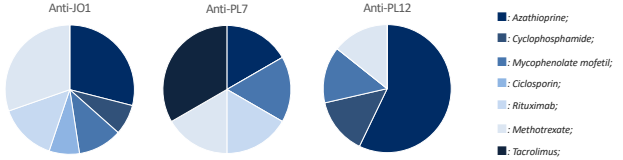
Figure 2 : Initial HRCT from patient with anti-PL7

Initial PFT

	Anti-JO1 (n = 16)	Anti-PL12 (n = 3)	Anti-PL7 (n = 5)
TLC, % pred	73,3 ± 28,6 [32 - 122]	81,3 ± 6,4 [74 – 86]	75 ± 14,3 [58 - 95]
VC, % pred	73,7 ± 22,3 [35 - 109]	78,7 ± 5,9 [72 - 83]	68,4 ± 11,4 [54 - 81]
FVC, % pred	69,3 ± 19,6 [42 - 109]	79 ± 6,9 [71 - 83]	72,40 ± 13,7 [53 - 87]
FEV_1, % pred	70,1 ± 18,4 [41 - 101]	74,7 ± 6,8 [67 - 80]	75,6 ± 11,1 [60 - 88]
FEV_1/FVC	81,7 ± 10,5 [59 - 100]	80,8 ± 3,7 [76,6 - 83]	86,5 ± 18 [59 - 108]
DLCO, % pred	49,7 ± 19,6 [18 - 82]	58,3 ± 14,6 [42 - 70]	41,8 ± 6,8 [32 - 50]
DLCO/VA, % pred	79,9 ± 21,3 [45 - 121]	78,7 ± 16,2 [60 - 89]	66,6 ± 22 [41 - 97]

Data are expressed as "mean ± standard deviation [range]"

Corticoid sparing immunosuppressive agents



CONCLUSION

- ASS exhibit variable radiological features and demographic characteristics.
- ILD mainly drives the progonosis of ASS and should be monitorised closely by regular PFT and HRCT.
- Treatment management is still challenging and must be closely monitorized. Further longitudinal clinical investigation are higly needed.