

ILD 多專科討論會

日期：2020/04/30 09:00-10:00

地點：第一醫療大樓 4 樓內科部會議室

No.	姓名	病歷號	收案日期	VS	風濕科診斷	影像學診斷
1	叢成玲	501836A	2019/6/17	謝祖怡	SLE with RA	ILD, UIP should be considered
2	賴苡岑	1242140I	2020/3/2	洪維廷	Sjogren's syndrome	NSIP pattern Progressive change
3	邱法準	2754954I	2019/10/25	林靖才	Sjogren's syndrome	Definite UIP
4	陳朝森	2562986B	2019/3/18	林靖才	Sjogren's syndrome	CPFE with probable UIP

FVC	1.47 (52%)	1.36 (50%)	1.33 (48%)	1.41 (51%)
FEV1	1.19 (52%)	1.11 (49%)	1.09 (48%)	1.16 (50%)
FEV1/FVC	81 %	81%	82%	82%
FEF 25-75%	1.27 (50%)	1.15 (46%)	1.14 (45%)	1.26 (50%)
TLC	2.33 (52%)	2.25 (52%)	2.67 (60%)	2.29 (51%)
DLCO	7.73 (51%)	6.58 (44%)	7.52 (50%)	6.04 (40%)

☐ **HRCT [2020/02/26]**

Finding:

Non-contrast CT scan of the chest showed:

Patient did not receive contrast medium administration. Tissue perfusion and vascular patency could not be evaluated and subtle lesion might be under estimated.

> Reticulation over periphery of right lower lobe and lower lobe with honeycombing pattern, interstitial lung disease, UIP should be considered. Suggest clinical correlation. No obvious change as compared with previous CT on 20190328

> Some small LNs in middle mediastinum. > No enlargement of bil. adrenal glands.

> No obvious air-trapping noted in the expiratory phase.

> DJD change of T-L spine with spur formation.

Impression:

1. Stable lung condition as compared with previous CT on 20190328.

2. For other details and DDx, please seen above description.

☐ **Cardiac ultrasound [2019/07/12]**

CONC. LVH (1.1, 1.1 CM)

AORTIC ROOT DILATATION (3.8 CM) WITH MILD AR

PROLAPSE OF ANTERIOR MITRAL LEAFLET WITH MILD MR

MILD TR WITH PEAK SYSTOLIC PRESSURE GRADIENT -- **20 MMHG**

DILATED PA TRUNK (-- 2.8 CM) WITH MILD PR REVERSE MV E/A RATIO

THE LV EJECTION FRACTION IS **53 %** NORMAL LV SYSTOLIC WALL MOTION.

☐ **討論事項及結論：■**

1. 是否為 **ILD** ? ☐是 ☐否
2. 是否為 **Indeterminate** ? ☐是 ☐否
3. 是否為 **UIP** ? ☐是 ☐否
4. 是否還有 **NSIP pattern** ? ☐是 ☐否
5. 是否還有免風疾病活動性(**activity**) 病變 ? ☐是 ☐否
6. 是否 **ILD** 持續進展 ? ☐是 ☐否
7. 是否調整免疫治療藥物 ? ☐是 ☐否
8. 是否建議使用抗肺纖維化藥物 ? ☐是 ☐否

No.	姓名	病歷號	收案日期	VS	風濕科診斷	影像學診斷
2	賴苡岑	1242140I	2020/3/2	洪維廷	Sjogren's syndrome	NSIP pattern Progressive change

□ Brief case summary

This is a 28 y/o female with 3 years history of Sjogren's syndrome, characterized by dry eye, dry mouth, positive Schirmer's test, sialoscintigraphy, SSA(52KD). Skin hyperpigmentation suspected HCQ related was noted later. She came to IMRH OPD for second opinion since 2018. Proximal muscle weakness and elevated CK were noted during followed up, dermatomyositis was suspected. Myositis survey was arranged but negative finding(EMG/NCV or muscle scan), while interstitial lung disease was noted. She accepted MTP pulse 400mg*6(2019/01,2019/07), rituximab 500mg*3(2019/07-08, 2020/02), MTP pulse 500mg*3(2020/02) for relapsing elevated CK and NSIP. HRCT showed progressive NSIP in 2020/02. MTP pulse was suggested but patient refused for fear of COVID-19. Oral Endoxan was prescribed since 2020/03.

Current medication:

- Bronchodilator: NEXThaler Foster(FormoteroL6+Beclomethasone100mcg) BID
- Immunosuppressive agent: CYC 50mg Q2D, Metholone tab 16mg(PD 0.76mg/kg/D)
- Anti-fibrotic agent: nil
- Pulmonary hypertension agent: nil
- Others: HCQ 200mg QD, montelukast 10mg HS

□ Laboratory

Immunologic profile (2020/03/02)		Biologic markers in ILD	
ANA	Negative	Ferritin	1579.42 (2020/02/03)
SS-A	> 240	ESR	21 (2020/03/02)
SS-B	Negative	hs-CRP	5.241(2020/02/03)--> 0.145(2020/03/02)
RF	Negative	CA-199	26.70
Scl-70	Negative	CA-153	34.04
Myositis Ab	Ro-52:+++	CA-125	32.90
Jo-1	Negative	NT-ProBNP	116.00
		6MWT	已預約排程

□ Pulmonary function test

日期	2020/02/03	2020/03/02
FVC	0.91 (35%)	1.25 (49%)
FEV1	0.86 (37%)	1.14 (49%)
FEV1/FVC	95 %	91%
FEF 25-75%	1.27 (43%)	1.50 (51%)

TLC	2.00 (59%)	NA
DLCO	NA	6.08 (42%)

❑ **HRCT [2020/02/12]**

Finding:

HRCT with patient in supine position, without contrast enhancement and with both inspiration phase and expiration phase, in axial and coronal reformation, shows:

- > Patches of mixed reticular and ground glass opacities over basal portion of RLL and LLL, increased the involvement lung parenchyma and extending to subpleural region, the picture could be compatible with NSIP pattern and suggest active inflammatory process.
- > Mild traction bronchiectasis over basal portion of RLL and LLL.
- > Diffuse centrilobular small ground glass nodular opacities of both lungs with some cluster over subpleural region, as a pattern of follicular bronchiolitis. In favor of inflammatory foci of both lungs.
- > No air trapping.
- > The diameter of main pulmonary artery is larger than aorta at the same level(Ser3,Img15), C/W pulmonary hypertension.
- > Prominent spleen.
- > No enlarged mediastinal node.
- > Otherwise, no other significant finding.

Impression:

- > C/W NSIP pattern. Progressive change is noted as compared with 2018/08/27.
- > C/W pulmonary hypertension.

❑ **Cardiac ultrasound [2019/01/07]**

PROLAPSE OF ANTERIOR MITRAL LEAFLETS WITH MILD MR

MILD TR WITH PEAK/MEAN SYSTOLIC PG -- **42/29 MMHG**

SUGGEST PULMONARY HYPERTENSION

MILD AR, MINIMAL PR

NORMAL LV SYSTOLIC WALL MOTION.

THE LV EJECTION FRACTION IS **58 %** MV E/A RATIO > 1

❑ **討論事項及結論：**

1. 是否為 **ILD** ? ☐是 ☐否
2. 是否為 **Indeterminate** ? ☐是 ☐否
3. 是否為 **UIP** ? ☐是 ☐否
4. 是否還有 **NSIP pattern** ? ☐是 ☐否
5. 是否還有免風疾病活動性(**activity**) 病變 ? ☐是 ☐否
6. 是否 **ILD** 持續進展 ? ☐是 ☐否
7. 是否調整免疫治療藥物 ? ☐是 ☐否

8. 是否建議使用抗肺纖維化藥物？☐是 ☐否

No.	姓名	病歷號	收案日期	VS	風濕科診斷	影像學診斷
3	邱法準	2754954I	2019/10/25	林靖才	Sjogren's syndrome 無免風科重大傷病 有肺癌,甲狀腺癌,膀胱癌重大傷病	Definite UIP

□ Brief case summary

This is a 57 y/o male with bladder cancer(s/p operation, high grade, cT1), thyroid cancer(pT4bN1bM1, stage IVB), lung cancer(s/p operation, cT1c) and 3 years history of Interstitial lung disease, characterized by PIPs, DIPs arthritis, dyspnea, positive ANA(fine speckled), RF(113.5 IU/mL), SSA(52KD). He came to CMUH first, HCQ BID and Myfortic 180mg BID were given. He came to IMRH OPD for second opinion in 2019/09. HRCT showed Definite UIP. For poor lung function, self funded Ofev(nintedanib) was prescribed since then. No data for anti-CCP, Schirmer's test or sialoscintigraphy.

Smoking history: ex-smoker, 1PPD for 15 years, quit since 1996.

Current medication:

- Bronchodilator: Spiolto(Tiotropium/OlodateroL 2.5/2.5mcg) QD
- Immunosuppressive agent: nil
- Anti-fibrotic agent: Ofev(nintedanib) 150mg BID
- Pulmonary hypertension agent: nil
- Others: HCQ 200mg BID

□ Laboratory

Immunologic profile		Biologic markers in ILD																																																									
ANA	1:160	Ferritin	182.31	(2020/03/16)																																																							
SS-A	61	ESR	11	(2020/03/16)																																																							
SS-B	Negative	hs-CRP	0.263	(2020/03/16)																																																							
RF	113.5	CA-199	5.44	(2020/03/02)																																																							
Scl-70	Negative	CA-153	23.32	(2020/03/02)																																																							
Myositis Ab	Ro-52:+++	CA-125	37.41	(2020/03/02)																																																							
Jo-1	Negative	NT-ProBNP	16.91																																																								
		6MWT (2019/11/25)	<table><tr><td></td><td>MHR/THR</td><td>HR</td><td>SaO2</td><td>Distance</td><td>METs</td><td>O2 Cost</td><td>Borg Scale</td><td>O2 L/min</td></tr><tr><td>Pre</td><td></td><td>75</td><td>98</td><td></td><td></td><td>5</td><td>0</td><td></td></tr><tr><td>Post</td><td>163/130</td><td>121</td><td>92</td><td>525</td><td>3.8</td><td></td><td>2</td><td></td></tr><tr><td></td><td></td><td>FVC</td><td>FVC%</td><td>FEV1</td><td>FEV1%</td><td>FEV1/FVC(%)</td><td></td><td></td></tr><tr><td>PRE-EX</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/A</td><td></td><td></td></tr><tr><td>POST-EX</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/A</td><td></td><td></td></tr></table> <p>HR peak:121 spO2 peak:90%</p> <p>檢查結束反應大腿外側痠感約3分。</p>				MHR/THR	HR	SaO2	Distance	METs	O2 Cost	Borg Scale	O2 L/min	Pre		75	98			5	0		Post	163/130	121	92	525	3.8		2				FVC	FVC%	FEV1	FEV1%	FEV1/FVC(%)			PRE-EX	N/A	N/A	N/A	N/A	N/A	N/A			POST-EX	N/A	N/A	N/A	N/A	N/A	N/A		
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❑ Pulmonary function test

日期	2019/10/25	2020/02/17
FVC	2.67 (61%)	2.62 (60%)
FEV1	2.02 (57%)	2.18 (62%)
FEV1/FVC	76%	83%
FEF 25-75%	1.60 (44%)	2.34 (65%)
TLC	4.34 (66%)	4.18 (65%)
DLCO	15.60 (67%)	21.95 (94%)

❑ HRCT [2020/02/23]

Finding:

Without contrast enhanced HRCT of lung with both inspiratory phase and expiratory phase:

1. Reticular opacities and honeycombing with subpleural predominance and apicobasal gradient, traction bronchiectasis and bronchiolectasis with basilar lung predominance are noted. The image pattern might be definite usual interstitial pneumonia (UIP).
2. Multiple paraseptal emphysema and bullae in RLL are noted. Compared with previous CT on 2019/09/24, the lesions show progressing change.
3. Pleural thickening in the bilateral lower chest, especially R't side, is noted.
4. Dilatation of the esophagus and pulmonary artery are also noted.
5. According to the above finding, the possibility of progressive systemic sclerosis (PSS) interstitial lung disease (ILD) could not be excluded.
6. Some small lymph nodes in the pretracheal and subcarinal of mediastinum are noted.

Impression:

1. Definite UIP
2. Possibility of PSS-ILD could not be excluded, F/U

❑ Cardiac ultrasound [2019/10/30]

CONC. LVH (1.1, 1.1 CM)

AORTIC ROOT DILATATION (3.8 CM) WITH MINIMAL AR

DILATED PA TRUNK (-- 2.6 CM) WITH MILD PR
PROLAPSE OF ANTERIOR MITRAL LEAFLETS WITH MINIMAL MR
MILD TR WITH PEAK/MEAN SYSTOLIC PG -- **22/15 MMHG**
REVERSE MV E/A RATIO THE LV EJECTION FRACTION IS **56 %**
NORMAL LV SYSTOLIC WALL MOTION. MV E/E' SEPTAL — 11.61
MV E/E' LATERAL — 9.23

☐ 討論事項及結論：■

1. 是否為 **ILD** ? ☐是 ☐否
2. 是否為 **Indeterminate** ? ☐是 ☐否
3. 是否為 **UIP** ? ☐是 ☐否
4. 是否還有 **NSIP pattern** ? ☐是 ☐否
5. 是否還有免風疾病活動性(**activity**) 病變 ? ☐是 ☐否
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7. 是否調整免疫治療藥物 ? ☐是 ☐否
8. 是否建議使用抗肺纖維化藥物 ? ☐是 ☐否

No.	姓名	病歷號	收案日期	VS	風濕科診斷	影像學診斷
4	陳朝森	2562986B	2019/3/18	林靖才	Sjogren's syndrome	CPFE with probable UIP

□ Brief case summary

This is a 55 y/o male with 4 years history of COPD and Sjogren syndrome, characterized by institial lung disease with respiratory failure, s/p ETT+MV, positive SSA(60KD, 52KD), Schirmer test, Saxon test. Followed up 2D echo showed pulmonary hypertension(51/42mmHg) in 2019. Revatio was applied. Followed up HRCT showed combined pulmonary fibrosis and emphysema(CPFE) with probable UIP and disease progression.

Smoking history: ex-smoker, 3-4PPD for 20+ years, quit since 2007.

Current medication:

- Bronchodilator: Spiolto(Tiotropium/OlodateroL 2.5/2.5mcg) QD
- Immunosuppressive agent: Metisone 4mg QD
- Anti-fibrotic agent: nil
- Pulmonary hypertensionagent: Revatio(Sildenafil) 20mg TID
- Others: HCQ 200mg QD

□ Laboratory

Immunologic profile (2020/03/13)		Biologic markers in ILD																																																																
ANA	1:1280	Ferritin	139.27																																																															
SS-A	> 240	ESR	15																																																															
SS-B	> 320	hs-CRP	1.136																																																															
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□ Pulmonary function test

日期	2019/06/10	2019/09/02	2019/11/25	2020/02/17
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FVC	2.84 (80%)	2.98 (89%)	3.05 (91%)	2.90 (87%)
FEV1	1.95 (68%)	2.27 (83%)	2.05 (75%)	2.08 (76%)
FEV1/FVC	69 %	76%	67%	72%
FEF 25-75%	1.13 (37%)	1.78 (60%)	1.14 (39%)	1.37 (47%)
TLC	3.99 (72%)	4.63 (86%)	4.35 (82%)	4.56 (85%)
DLCO	5.92 (30%)	5.46 (28%)	5.53 (28%)	5.90 (30%)

❑ HRCT [2020/02/17]

Finding:

HRCT with patient in supine position, without contrast enhancement and with both inspiration phase and expiration phase, in axial and coronal reformation, shows:

1. CPFE with probable UIP.
2. More extensive fibrosis is noted esp. in basal portion of both lower lobe. Progressive change is considered.
3. Main PA measuring 3.47 cm. Suggest cardiac echo.
4. No enlarged mediastinal node.
5. Spot calcification of LAD.
6. Otherwise, no other significant finding.

Impression:

CPFE with probable UIP.

Progression of disease as compared with 2019/02/20.

❑ Cardiac ultrasound [2020/03/16]

LA(4.5), RV(3.2) CHAMBER DILATATION

CONC. LVH (1.1, 1.2 CM)

AORTIC VALVE THICKENED WITH MILD AR

PROLAPSE OF ANTREIOR MITRAL LEAFLETS WITH MILD MR

MILD TR WITH PEAK/MEAN SYSTOLIC PG -- **43/27 MMHG**

DILATED PA TRUNK (-- 2.7 CM) WITH MINIMAL PR

NORMAL LV SYSTOLIC WALL MOTION.

THE LV EJECTION FRACTION IS **64 %**

REVERSE MV E/A RATIO MV E/E' SEPTAL— 4.2 MV E/E' LATERAL — 3.9

❑ 討論事項及結論：■

1. 是否為 **ILD** ? ☐是 ☐否
2. 是否為 **Indeterminate** ? ☐是 ☐否
3. 是否為 **UIP** ? ☐是 ☐否
4. 是否還有 **NSIP pattern** ? ☐是 ☐否

- 5. 是否還有免風疾病活動性(activity) 病變？☐是 ☐否
- 6. 是否 ILD 持續進展？☐是 ☐否
- 7. 是否調整免疫治療藥物？☐是 ☐否
- 8. 是否建議使用抗肺纖維化藥物？☐是 ☐否