Pleuroparenchymal fibroelastosis in UIP CT pattern: our experiece

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Case report 1 04/2017

Wom… …ucasian
Admi…

Risk f…
Form…
Non s…
Slight…

Comorbidities:
Nodular tireopathy, GE reflux

Symp…
Exerti… …onths and
dry c…

Physi…
No Ra… …tal
clubb…
Bi-bas…

Blood exams

- Autoimmunity screening: negative
- CA 15.3= 48.4 U/ml
- CA 19.9=176.8 U/ml
- NSE= 20.8 ng/ml
- Vitamin D =7.3 ng/ml

Fiberoptic bronchoscopy (BAL)

- Total cells x10^5/ml
- Neutrophils 53% (1-2%)
- Eosinophils 7% (0-0.5%)
- Macrophages 35% (75-85)
- Lymphocytes 5% (8-12)
- CD4/CD8: 0.8 (0.8-2)

Blood gases analysis at rest in ambient air
pO_2 8…
pCO_2…
PH 7.…
HCO_3…

Spiro…
FVC 1…
FEV_1…
IT 96.…
TLC 1.88 L (60% pred)

CO d…
DLC(… …red)
DLC(… …red)

6MV…
Test… …spnoea
Initi…
Dist…

Echocardiography

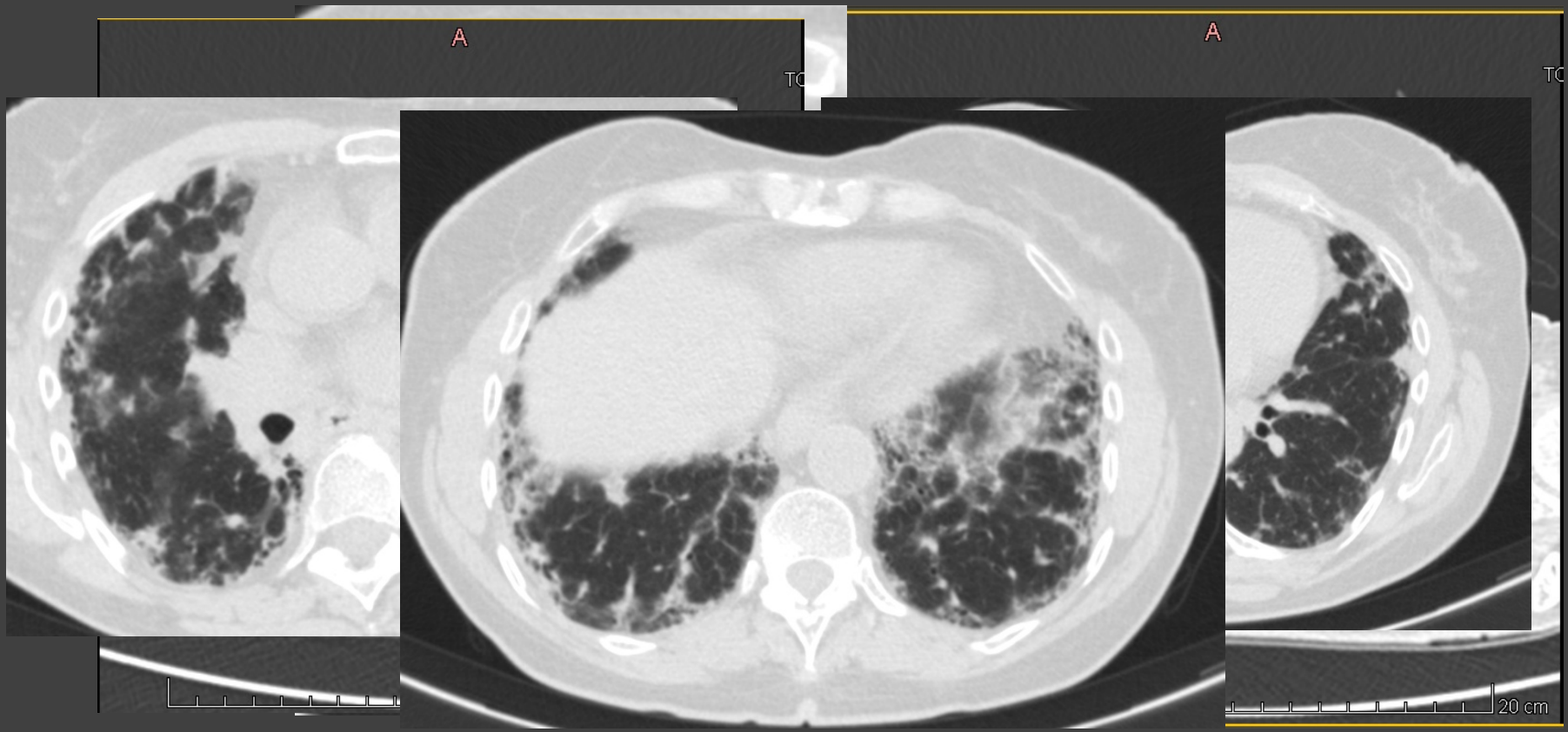
- EF 60%
- TAPSE 2.8 cm
- Estimated systolic PAP 35 mmHg

Nocturnal cardio-respiratory testing

- positive
- AHI 9.2/h
- ODI 11.1/h
- t90 0%

E.C.

HRCT 2017



Courtesy of Dott. G. Rea, Radiology Monaldi Hospital

Management

Criobiopsy was not indicated as thorax HRCT was diagnostic of UIP definite

Lung transplantation (LT) not indicated due to low bio-umoral conditions + family reasons

MDD: Idiopathic pulmonary fibrosis with associated CT findings highly suggestive of PPFE

Bridge therapy with an anti-fibrotic drug and respiratory rehabilitation on hold better bio-umoral conditions for LT

Infection prevention (vaccination)

Nutritional status assessment and correction of Vitamin D deficiency

Oxygen supplementation

GE reflux therapy

Clinical and functional monitoring every three months

Case report 2.08/2017

Wo , Caucasian
Tip

Risk
No
Act
Ato

Comorbidities:
Prev therapy
due

Sym
Chr

Phy
No al clubbing
Bi-b

Blood exams

- Autoimmunity screening: negative
- Tumoral markers screening: negative
- Vitamin D = 6.1 ng/ml

Fiberoptic bronchoscopy (BAL)

- Total cells $7.7x10^6$/ml
- Neutrophils 23% (1-2%)
- Eosinophils 4% (0-0.5%)
- Macrophages 71% (75-85)
- Lymphocytes 2% (8-12)
- CD4/CD8: 1.2 (0.8-2)

Blood gases analysis at rest in ambient air

pO_2 77 mmHg
pCO_2
PH 7
HCO

Spiro
FVC
FEV_1
IT 92.1%
RV 0.91 L (56% pred)
TLC 2.38 L (49% pred)

CO
DLC pred)
DLC pred)

6M
Test
dys
Init
Dist

Echocardiography

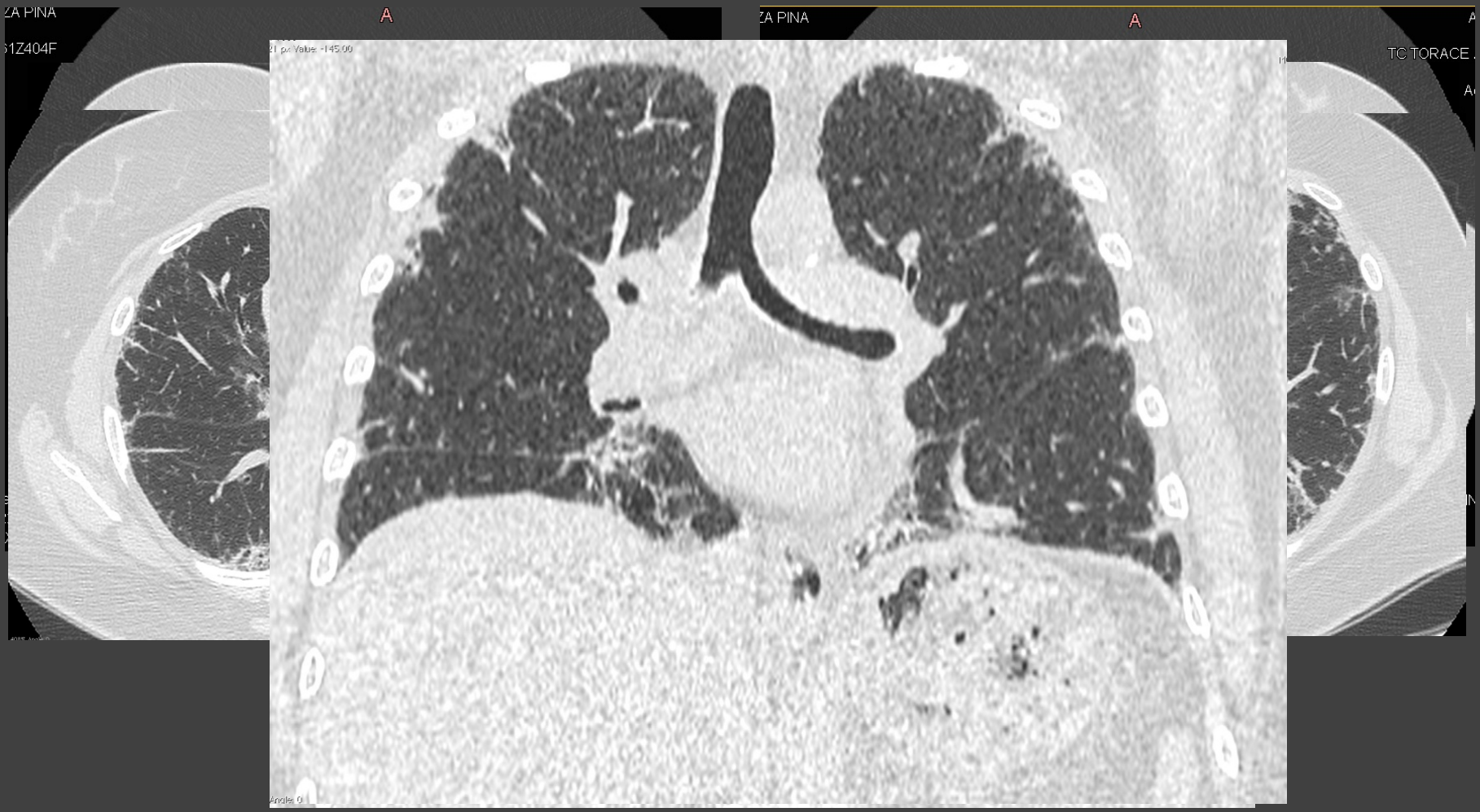
- EF 60%
- TAPSE 2.0 cm
- Estimated systolic PAPs 20 mmHg

Nocturnal cardio-respiratory testing

- negative
- AHI
- ODI
- t90

F.V.

HRCT 2017



Courtesy of Dott. G. Rea, Radiology Monaldi Hospital

Management

Criobiopsy should be considered with thorax HRCT diagnostic of UIP possible (probable)

Indication of lung transplantation

MDD: Idiopathic pulmonary fibrosis with associated CT findings highly suggestive of PPFE

Weight correction

Pulmonary rehabilitation

Bridge therapy with an anti-fibrotic drug

Infection prevention (vaccination)

Nutritional status assessment and correction of Vitamin D deficiency

Oxygen supplementation

GE reflux therapy

Clinical and functional monitoring every three months

Case

Man, sian
Post

Risk f
No sr
Activ nany
yrs

Como
GE re
Atria

Symp
Exert nths
and c

Physi
No R bbing
Bi-basal inspiratory crackles

Blood exams

- Autoimmunity screening: negative
- Tumoral markers screening:
- CA-15.3=29 U/l)
- CA-125=94.7 U/l)
- CA-19.9=101.5 U/l
- Vitamin D = 19.6 ng/ml

Fiberoptic bronchoscopy (BAL)

- not performed

Blood gase ir
pO_2 60 mn
pCO_2 39 m
PH 7.43
HCO_3^- 26.4

Spirometr
FVC 1.21 L
FEV_1 1.16 l
IT 95.8%
RV 0.68 L (26% pred)
TLC 2.10 L

CO diffusin
$DLCO_{sb}$ 4.1
$DLCO_{VA}$ 2.0

6MWT
Test stoppe noea
Initial O_2 Sa
Distance walked 99 m (35.8% pred)

Echocardiography

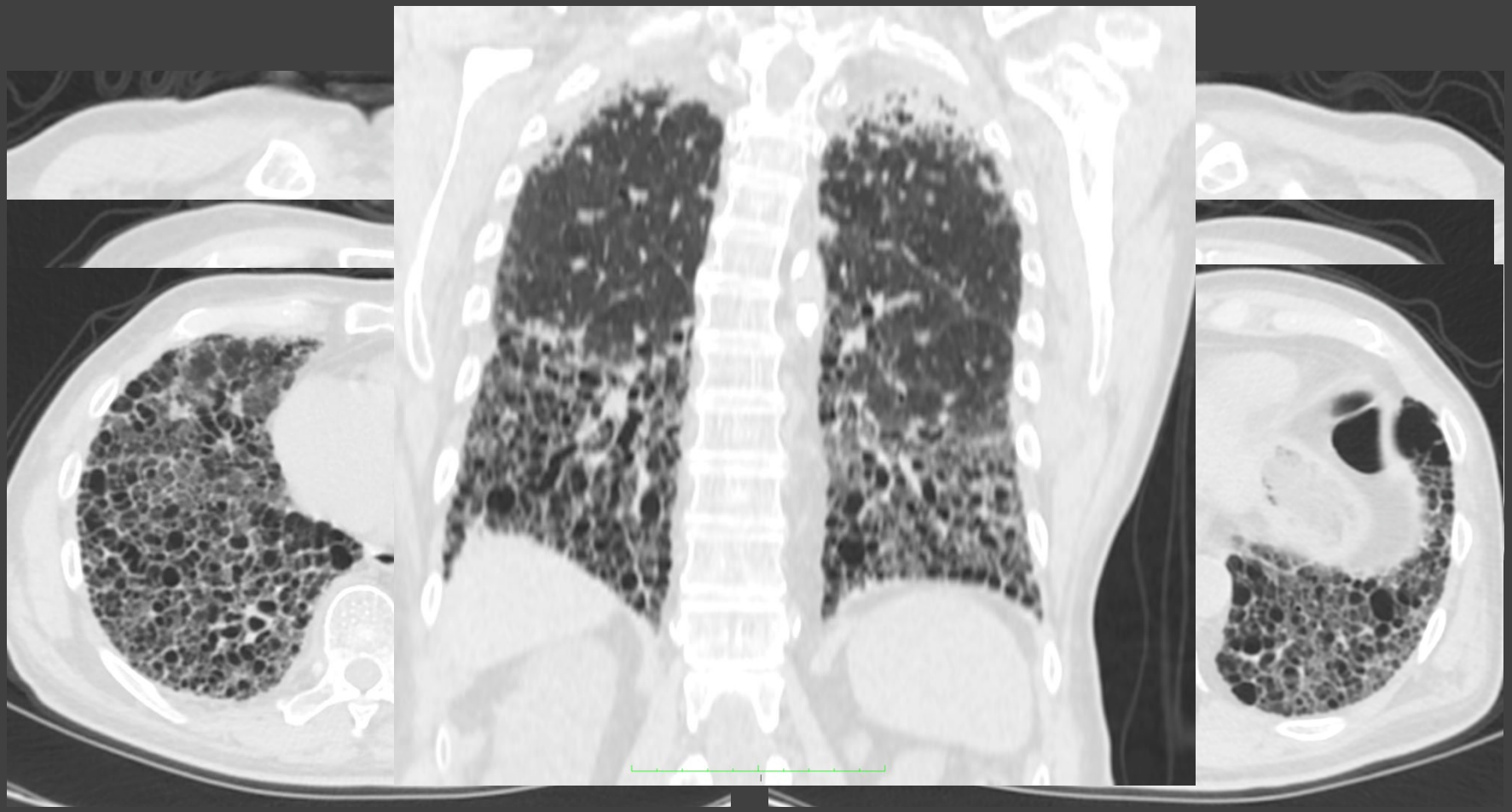
- EF 55%
- TAPSE 18 mm
- Estimated systolic PAP within the limits

Nocturnal cardio-respiratory testing

- negative
- AHI 1.9/h
- ODI 6.8/h
- T90 16%

HRCT 2018

R.G.



Courtesy of Dott. G. Rea, Radiology Monaldi Hospital

Management

Thorax HRCT was diagnostic of UIP definite
Criobiopsy and lung transplantation were not indicated

MDD: Idiopathic pulmonary fibrosis with associated CT findings highly suggestive of PPFE

Pulmonary rehabilitation

Therapy with an anti-fibrotic drug was not indicated

Infection prevention (vaccination)

Correction of Vitamin D deficiency

Oxygen supplementation

GE reflux therapy

Clinical and functional monitoring every three months

What WAS wrong?

AMERICAN THORACIC SOCIETY DOCUMENTS

Diagnosis of Idiopathic Pulmonary Fibrosis
An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Jeffrey L. Myers, Luca Richeldi, Christopher J. Ryerson, David J. Lederer, Juergen Behr, Vincent Cottin, Sonye K. Danoff, Ferran Morell, Kevin R. Flaherty, Athol Wells, Fernando J. Martinez, Arata Azuma, Thomas J. Bice, Demosthenes Bouros, Kevin K. Brown, Harold R. Collard, Abhijit Duggal, Liam Galvin, Yoshikazu Inoue, R. Gisli Jenkins, Takeshi Johkoh, Ella A. Kazerooni, Masanori Kitaichi, Shandra L. Knight, George Mansour, Andrew G. Nicholson, Sudhakar N. J. Pipavath, Ivette Buendía-Roldán, Moisés Selman, William D. Travis, Simon Walsh, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society

This official clinical practice guideline of the American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society (JRS), and Latin American Thoracic Society (ALAT) was approved by the ATS, JRS, and ALAT May 2018, and the ERS June 2018

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distinct from IPF/UIP and may well be classified as PPFE.

CHEST NOVEMBER 2014

Open questions

Is the CT pattern of PPFE/UIP representative of a disease entity distinct from IPF in some cases?

Is such a pattern representative of a distinct phenotype of IPF (more aggressive)?

Should the combination of PPFE/UIP be considered as the pathological background of a combined syndrome?

Are we going to treat with new anti-fibrotic drugs patients with a fibrotic CT pattern until lung transplantation as having IPF?

Thank you all for your attention