PNEUMOMEDICINA 2022

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I dubbi del Dottor Watson

Alessandro Lucidi

Scuola di Specializzazione in Medicina Interna
Università degli Studi di Milano

ANAMNESI

Paziente di 66 anni, non intolleranze farmacologiche note, ex bracciante agricola.

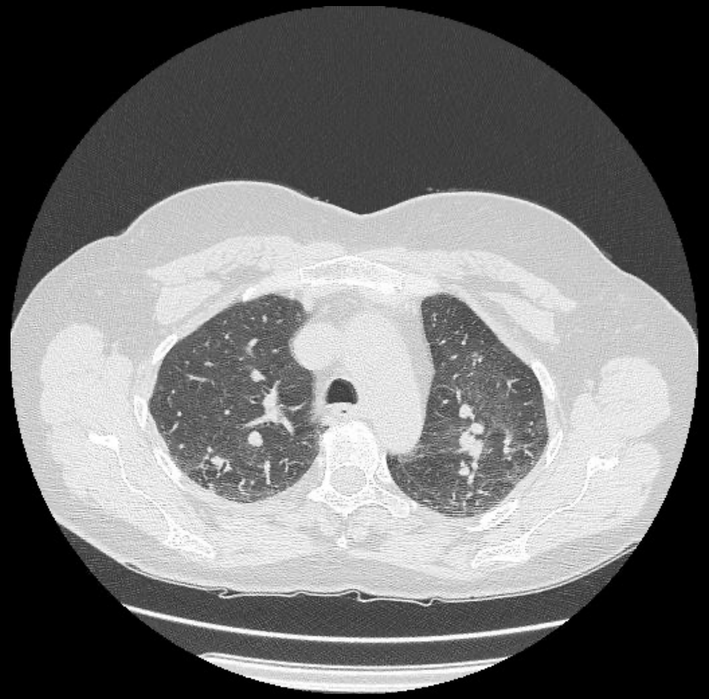
Non storia di tabagismo.

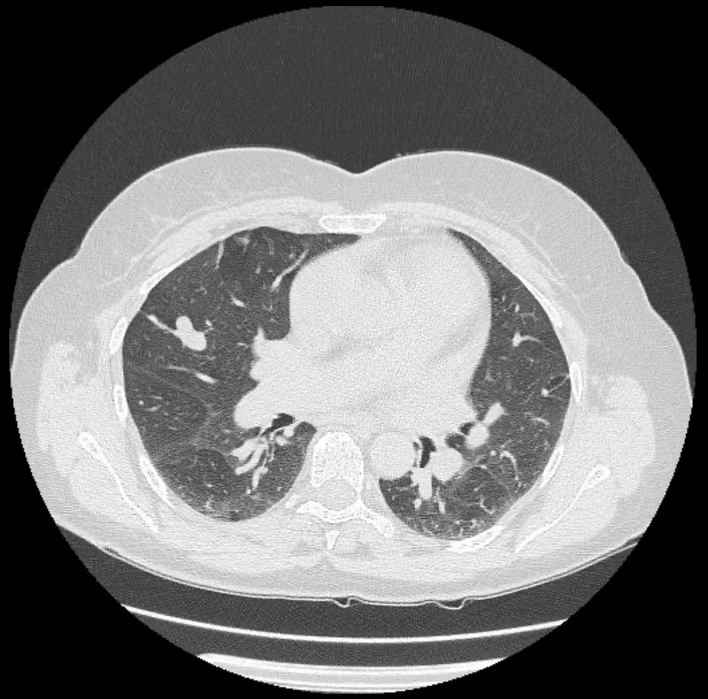
In anamnesi patologica remota: ipertensione arteriosa essenziale
cardiopatia scleroipertensiva.

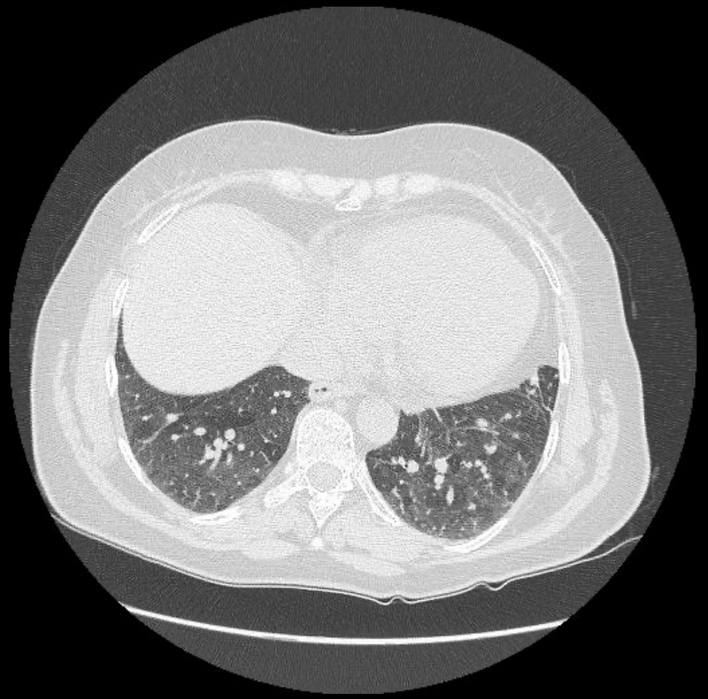
STORIA PNEUMOLOGICA

A Febbraio 2019 ricovero c/o altro Ospedale per broncopolmonite destra e dispnea da sforzo.

La paziente giungeva in visita ambulatoriale ad Aprile 2019. Persisteva dispnea da sforzo. Eseguita TC del torace.







TC torace: plurime consolidazioni pseudonodulari parenchimali bilaterali a distribuzione peribronchiale (la maggiore al LM). Nelle scansioni in espirio diffuse aree di air trapping.

A Giugno 2019 PET total body negativa a livello polmonare.

Quale indagine effettuare?

1. FBS
2. Esami ematici
3. Biopsia

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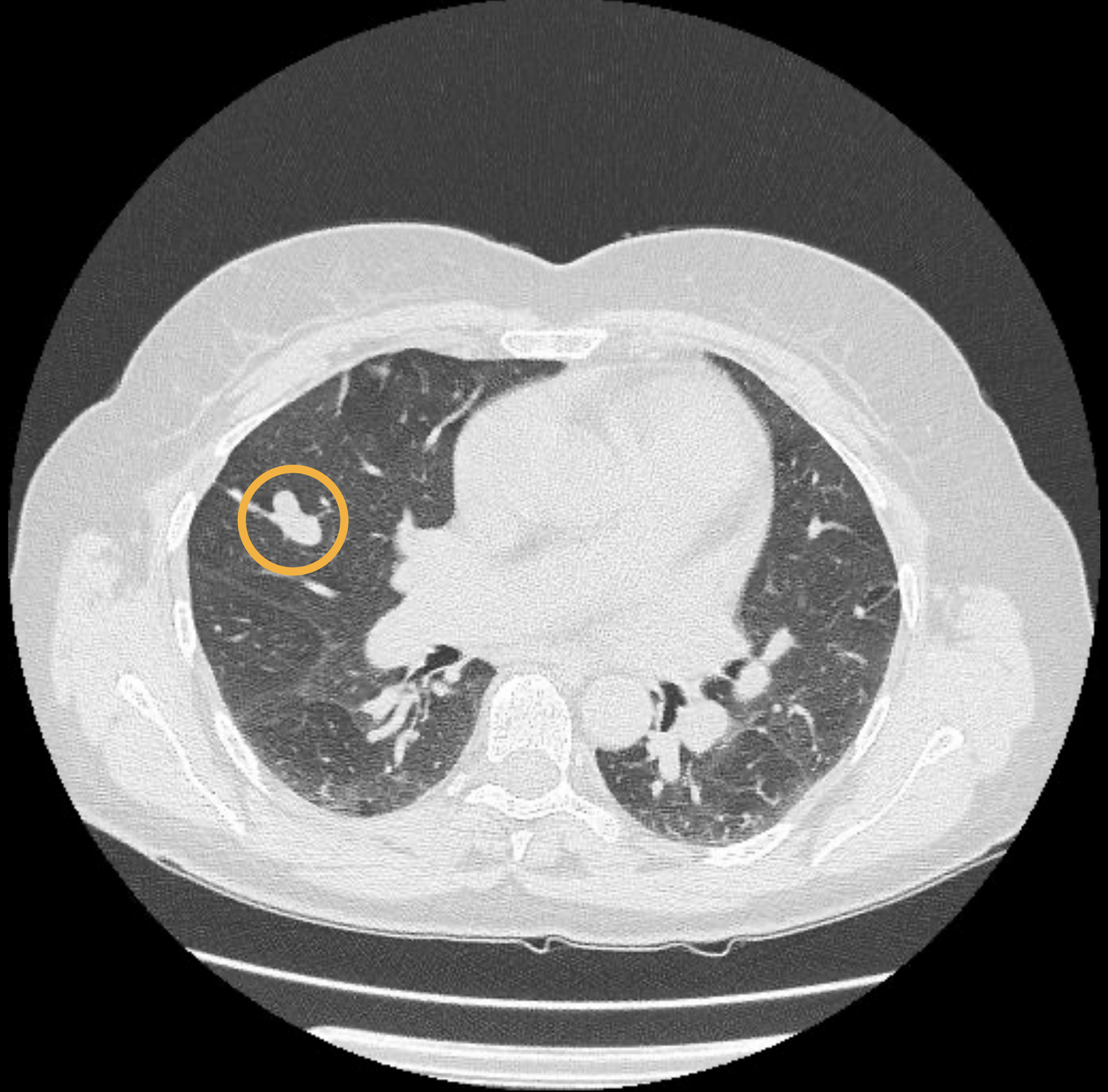
1. FBS
2. Esami ematici
3. Biopsia

Eseguita agobiopsia di: fibrosi stromale e focale infiltrato linfoide con alcuni granulomi a cellule epitelioidi e giganti plurinucleate senza necrosi.



Sospetto di sarcoidosi polmonare nodulare.

Elliot D. Crouser, et al. Diagnosis and Detection of Sarcoidosis An Official American Thoracic Society Clinical Practice Guideline 2020.

Screening di malattia sistemica:

1. Visita Oculistica: angiosclerosi retinica.
2. Funzione epatica e renale nella norma.
3. Metabolismo del calcio nella norma.

4. Ecocardiogramma: VS nella norma. Movimento anomalo del SIV per aumentate pressioni VD. Aneurisma destro convesso del SIA. Lieve IM. PAPs 52 mmHg.

Posta diagnosi di sarcoidosi con interessamento esclusivamente polmonare.

Quale terapia?

1. Solo steroide

2. Steroide e methotrexate

3. Steroide ed idrossiclorochina

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In pazienti con sarcoidosi polmonare iniziare con steroide orale.

Aggiungere methotrexate o altro immunosoppressore se malattia non controllata o effetti collaterali.

Baughman RP, Valeyre D, Korsten P, et al. ERS clinical practice guidelines on treatment of sarcoidosis. Eur Respir J 2021

A Luglio 2020 ricovero c/o altro Ospedale per second opinion.

1. TC torace: invariata rispetto alla precedente di Aprile 2019.

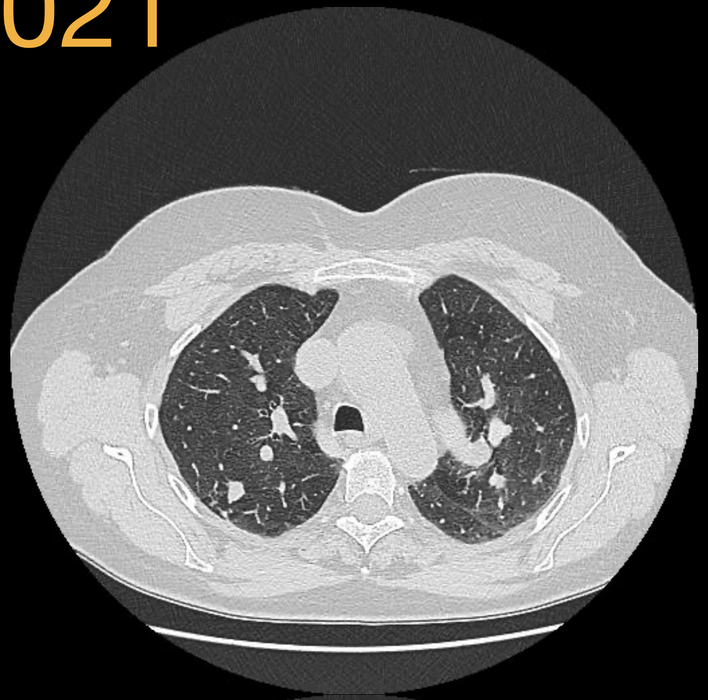
2. Broncoscopia: citologico su BAL negativo per CTM, microscopico BAL negativo per BAAR, PCR per BK su BAL negativa, colturale per BK negativo.

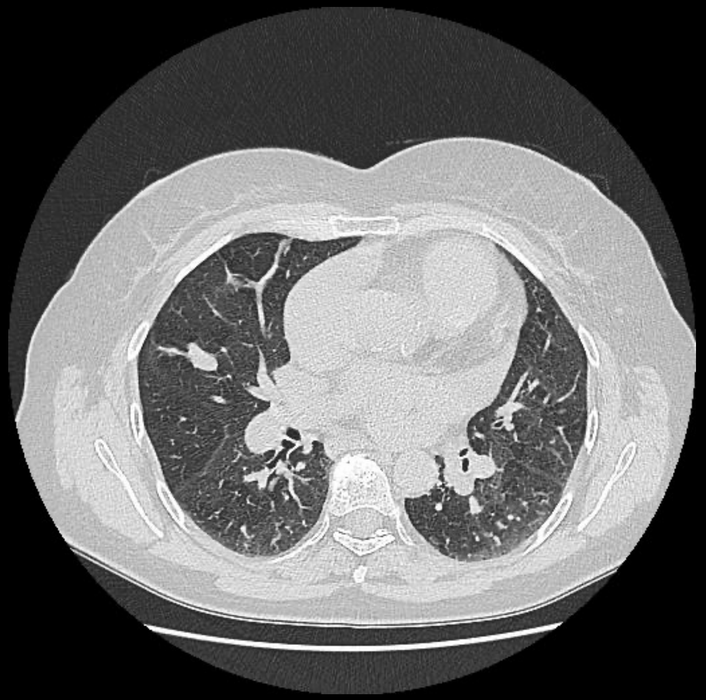
3. Citofluorimetria su BAL: macrofagi 89.5%, monociti 0%, granulociti 1.5%, linfociti 9%, eosinofili 0%.

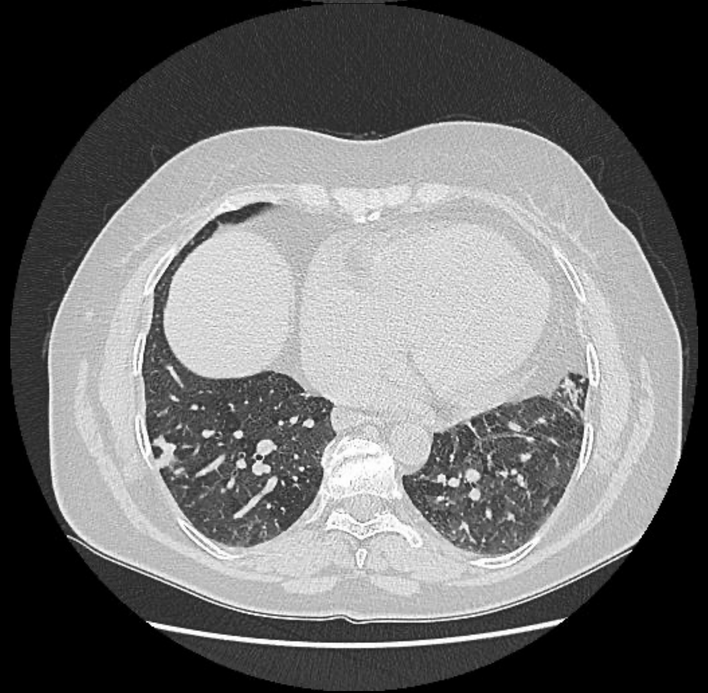
4. Ecocardiogramma: setto paradosso da sovraccarico destro, VD di dimensioni aumentate (46 x 48 mm), TRV 2.9 m/s, TAPSE 28 mm, PAPs 40 mmHg, aneurisma del SIA senza segni di shunt al Doppler.

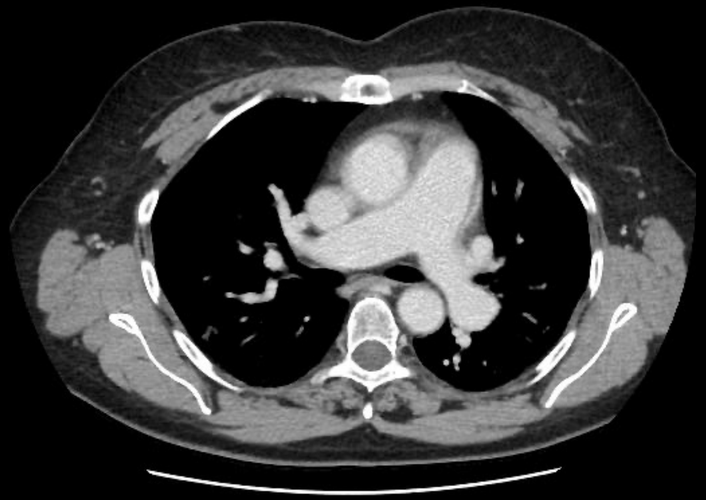
Vista la sostanziale asintomaticità e l’assenza di alveolite linfocitaria o coinvolgimento di altri organi si consigliava di interrompere la terapia steroidea.

Marzo 2021









Incremento volumetrico di 2 aree nodulariformi polilobulate al LSD e al LID. Non embolia polmonare.

RICOVERO

A Novembre 2021 ricovero c/o la nostra U.O. per rivalutazione e per peggioramento della dispnea da sforzo.

Cosa non è compatibile con la diagnosi di sarcoidosi?

Clinical Features Supportive of a Diagnosis of Sarcoidosis

	Highly Probable	Probable
History	Löfgren's syndrome*	Seventh cranial nerve paralysis
		Treatment-responsive renal failure
		Treatment-responsive CM or AVNB
		Spontaneous/inducible VT with no risk factors
Physical	Lupus pernio	Maculopapular, erythematous, or violaceous skin lesions
	Uveitis	Subcutaneous nodules
	Optic neuritis	Scleritis
	Erythema nodosum	Retinitis
		Lacrimal gland swelling
		Granulomatous lesions on direct laryngoscopy
		Symmetrical parotid enlargement
		Hepato-/splenomegaly
Imaging	Bilateral hilar adenopathy (CXR, CT, and PET)	Upper lobe or diffuse infiltrates (CXR, CT, and PET)
	Perilymphatic nodules (chest CT)	Peribronchial thickening (CT)
	Gadolinium enhancement on MRI (CNS)	Two or more enlarged extra thoracic nodes (CT, MRI, and PET)
	Osteolysis, cysts/punched-out lesion, trabecular pattern bone (X-ray, CT, and MRI)	Increased inflammatory activity in heart (MRI, PET, and gallium)
	Parotid uptake (gallium and PET)	Imaging showing enlargement or nodules in liver or spleen (CT, PET, and MRI)

Definition of abbreviations: ACE = angiotensin-converting enzyme; AV = atrioventricular; AVNB = atrioventricular node block; CM = cardiomyopathy; CNS = central nervous system; CT = computed tomography; CXR = chest X-ray; LVEF = left ventricular ejection fraction; MRI = magnetic resonance imaging; PET = positron emission tomography; VT = ventricular tachycardia.

*Löfgren's syndrome is defined as bilateral hilar adenopathy with erythema nodosum and/or periarticular arthritis.
†Abnormal vitamin D metabolism is defined as normal to low parathyroid hormone, normal to elevated 1,25-dihydroxyvitamin D, and normal to low 25-hydroxyvitamin D.
‡ACE elevated above 50% of the upper limit of normal was considered abnormal.

- Assenti aspetti clinici e radiologici caratteristici di sarcoidosi.

- Refrattarietà a terapia steroidea cronica.

- Sono state escluse tutte le altre possibili cause?

Elliot D. Crouser, et al. Diagnosis and Detection of Sarcoidosis
An Official American Thoracic Society Clinical Practice Guideline 2020.

Emogasanalisi arteriosa in aria ambiente: pH 7.45, pCO2 37.4 mmHg, pO2 84.9 mmHg, HCO3 25.9 mmol/L, SatO2 97.1%

PFR: FEV1 1.84 L (68%), FVC 2.25 L (65 %), Tiffenau 85%, TLC 4.79 L (78%), RV 2.29 L (78%), DLCO 20 mL/min/mmHg (84%).

Test del cammino: percorsi 450 m. Saturazione iniziale 97%, Saturazione finale 93%.

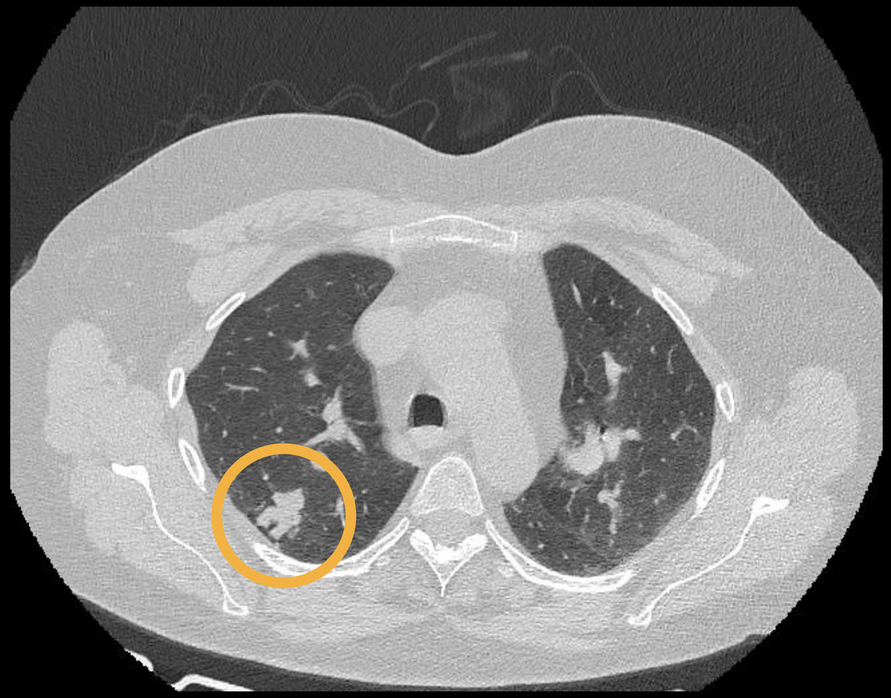
Ecocardiogramma: TRV 3.36 m/s, TAPSE 24 mm, PAPs 55 mmHg, appiattimento del SIV da sovraccarico ventricolare destro, ampio aneurisma del setto interatriale.

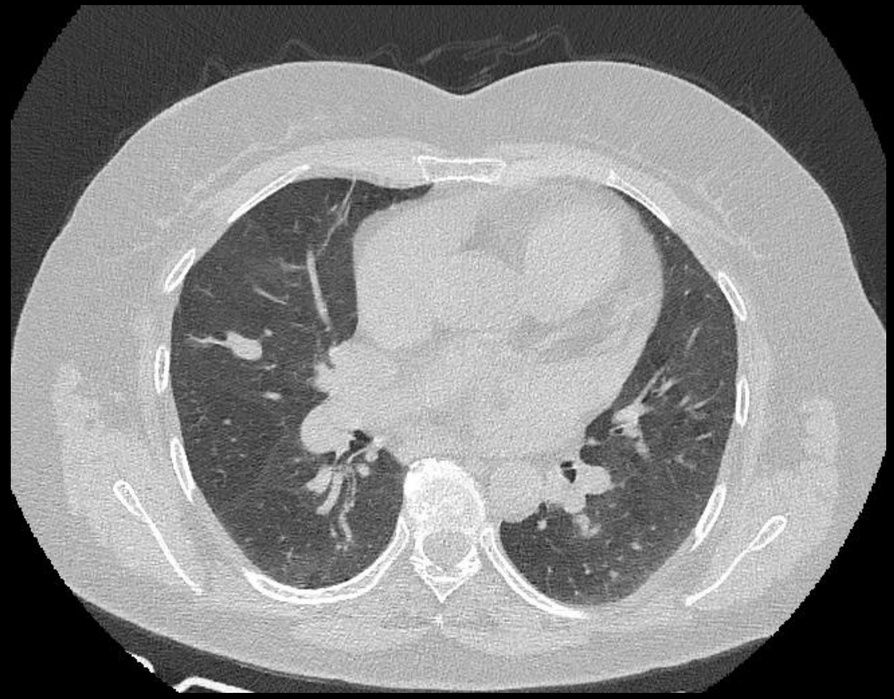
EcocolorDoppler trancranico: esame positivo per moderato shunt destro-sinistro

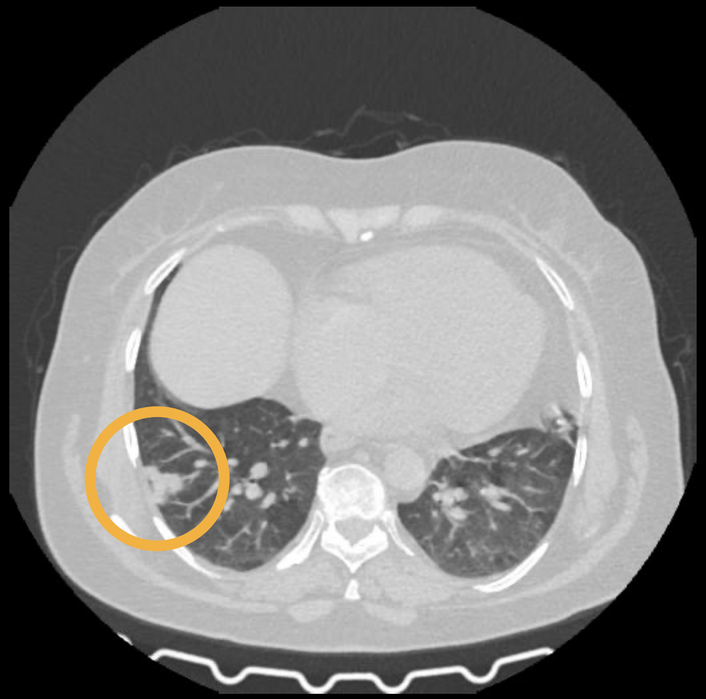
Ecocardiogramma transesofageo: presenza di DIA tipo OS con shunt interatriale bidirezionale.

Revisione preparati istologici relativi all’agobiopsia del 2019: processo infiammatorio nodulare linfocitario e granulomi di varie dimensioni privi di necrosi.

Compatibile con infiammazione granulomatosa cronica non altrimenti specificabile.

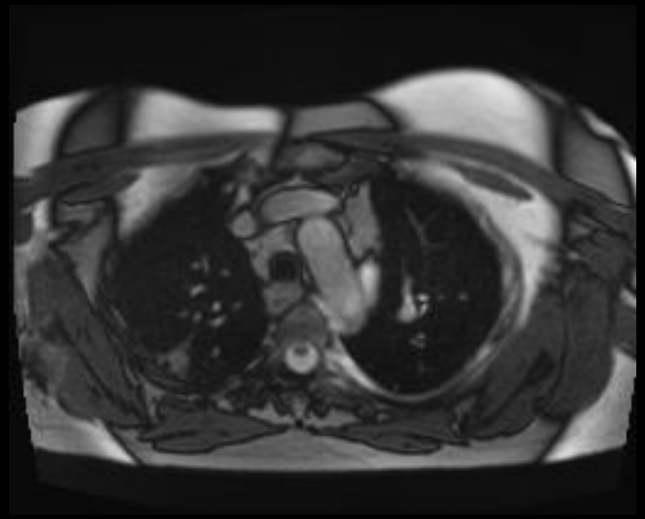


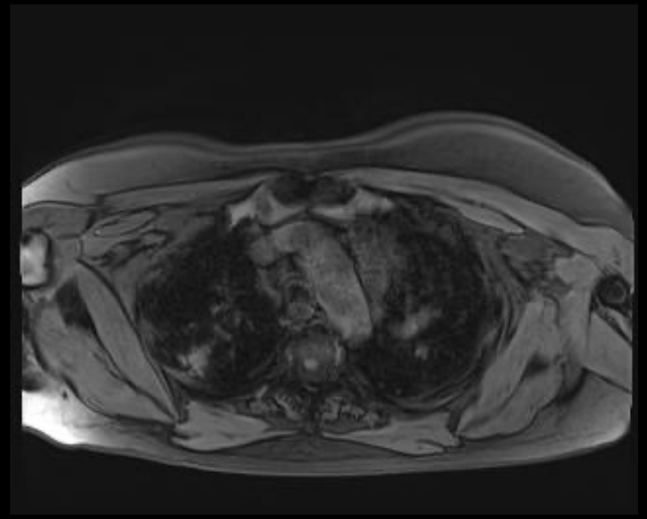


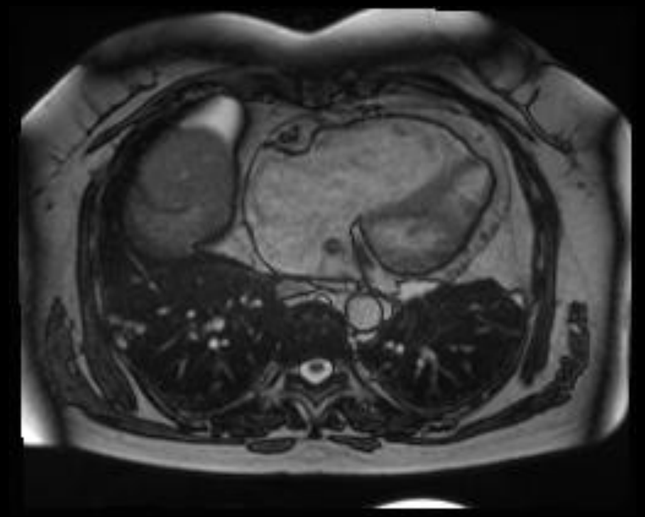


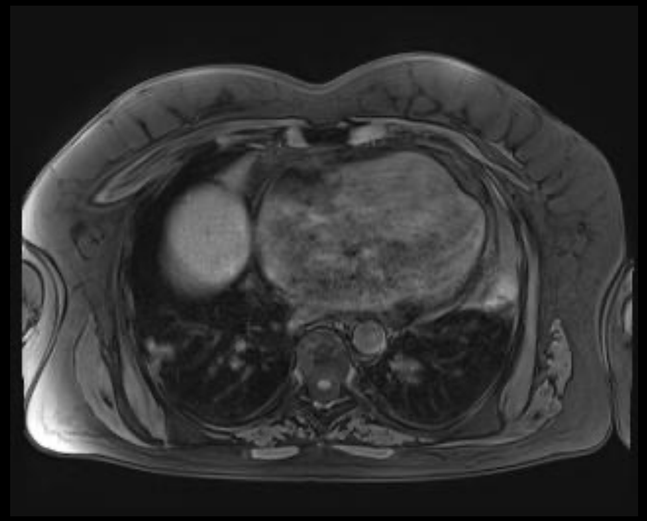
Opacità nodulari di aspetto ramificato, non calcificate né cavitate, a densità sovraidrica.

Discussione multidisciplinare.









RMN torace: Confermata natura mucoide.

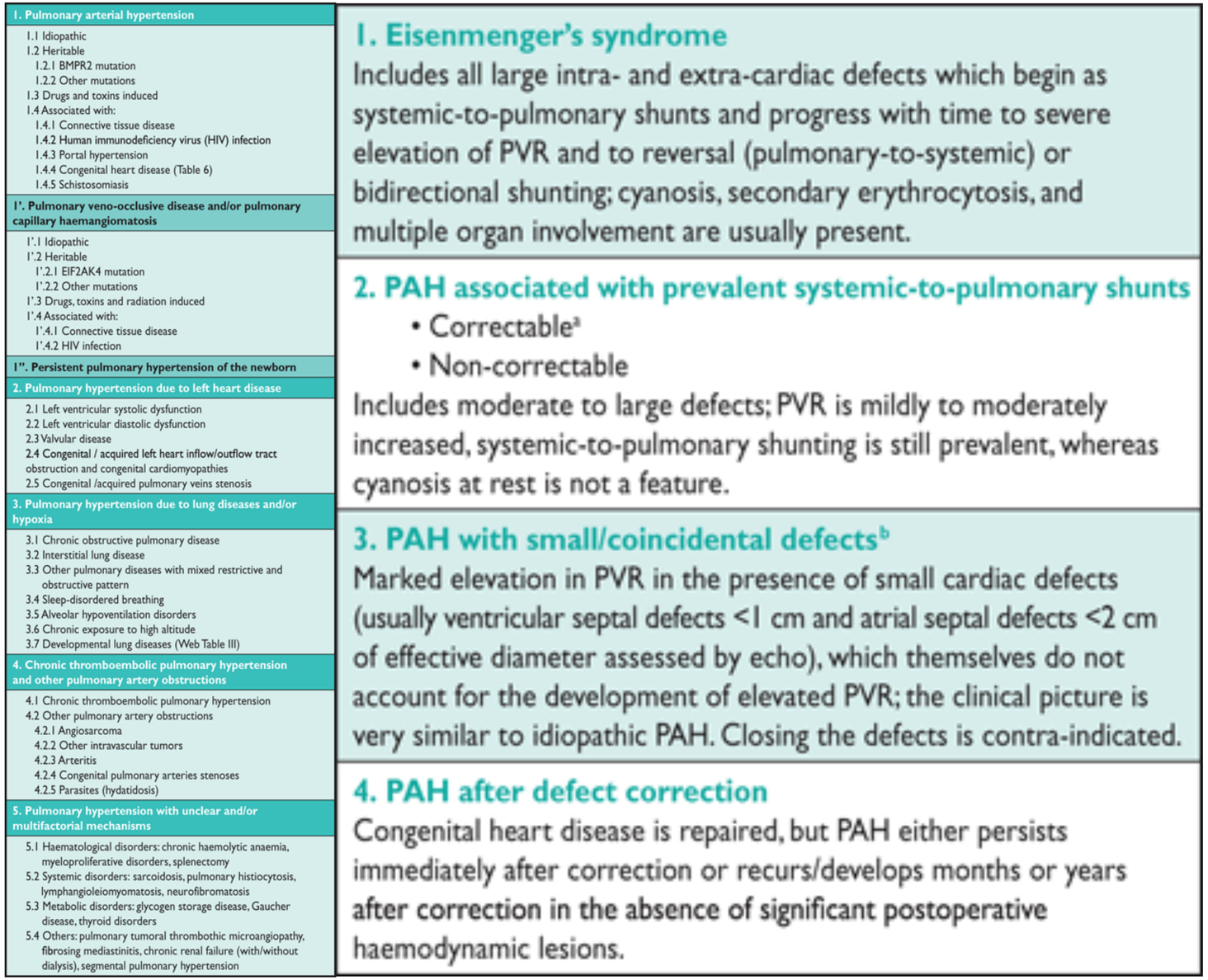
Si pone diagnosi di mucoid impaction.

IgE ed IgG specifiche per Aspergillus fumigatus ed IgE totali negative. Esclusa ABPA.

DIAGNOSI

Mucoid impaction in bronchiectasie distali.

Difetto del setto interatriale di tipo ostium secundum.



1. Pulmonary arterial hypertension
- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2 mutation
  - 1.2.2 Other mutations
- 1.3 Drugs and toxins induced
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 Human immunodeficiency virus (HIV) infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease (Table 6)
  - 1.4.5 Schistosomiasis

1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis
- 1'.1 Idiopathic
- 1'.2 Heritable
  - 1'.2.1 EIF2AK4 mutation
  - 1'.2.2 Other mutations
- 1'.3 Drugs, toxins and radiation induced
- 1'.4 Associated with:
  - 1'.4.1 Connective tissue disease
  - 1'.4.2 HIV infection

1''. Persistent pulmonary hypertension of the newborn

2. Pulmonary hypertension due to left heart disease
- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital / acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
- 2.5 Congenital /acquired pulmonary veins stenosis

3. Pulmonary hypertension due to lung diseases and/or hypoxia
- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental lung diseases (Web Table III)

4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions
- 4.1 Chronic thromboembolic pulmonary hypertension
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Angiosarcoma
  - 4.2.2 Other intravascular tumors
  - 4.2.3 Arteritis
  - 4.2.4 Congenital pulmonary arteries stenoses
  - 4.2.5 Parasites (hydatidosis)

5. Pulmonary hypertension with unclear and/or multifactorial mechanisms
- 5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis, neurofibromatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: pulmonary tumoral thrombotic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension

1. Eisenmenger's syndrome

Includes all large intra- and extra-cardiac defects which begin as systemic-to-pulmonary shunts and progress with time to severe elevation of PVR and to reversal (pulmonary-to-systemic) or bidirectional shunting; cyanosis, secondary erythrocytosis, and multiple organ involvement are usually present.

2. PAH associated with prevalent systemic-to-pulmonary shunts
- Correctable[a]
- Non-correctable

Includes moderate to large defects; PVR is mildly to moderately increased, systemic-to-pulmonary shunting is still prevalent, whereas cyanosis at rest is not a feature.

3. PAH with small/coincidental defects[b]

Marked elevation in PVR in the presence of small cardiac defects (usually ventricular septal defects <1 cm and atrial septal defects <2 cm of effective diameter assessed by echo), which themselves do not account for the development of elevated PVR; the clinical picture is very similar to idiopathic PAH. Closing the defects is contra-indicated.

4. PAH after defect correction

Congenital heart disease is repaired, but PAH either persists immediately after correction or recurs/develops months or years after correction in the absence of significant postoperative haemodynamic lesions.

DIA: inizialmente shunt sinistro-destro che nel tempo ha determinato ipertensione polmonare e successiva inversione dello shunt.

Galie` N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hyper- tension: the Joint Task Force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). Eur Respir J 2015; 46:903-75.