LUNGS AT WORK
www.lungsatwork.org.uk

Cases from the clinic

SW training day
9th October 2018

Johanna Feary

Dear Paul

Could I please ask for your advice about a 68 year old man who was referred because of a cough. He is not breathless. He has a long history of smoking but little else.

He worked as a carpenter and can recall no specific asbestos exposure.

He had a few harsh crackles at one base which led to a CT chest. There is very minor reticulation in the middle lobe, along with pleural plaques. Emphysema is the dominant feature.

Lung function is preserved: 2.4/3.9, being 90% and 110% predicted. TLCO 60%, KCO 75%.

I wouldn't be too worried about him – my only concern is that he appears to have (albeit limited) fibrosis in the context of no evident asbestos exposure.

Medicolegally, is this someone I should be advising to seek a legal opinion, or do you think that his lack of heavy exposure would make his having asbestosis unlikely?

An ILD doctor

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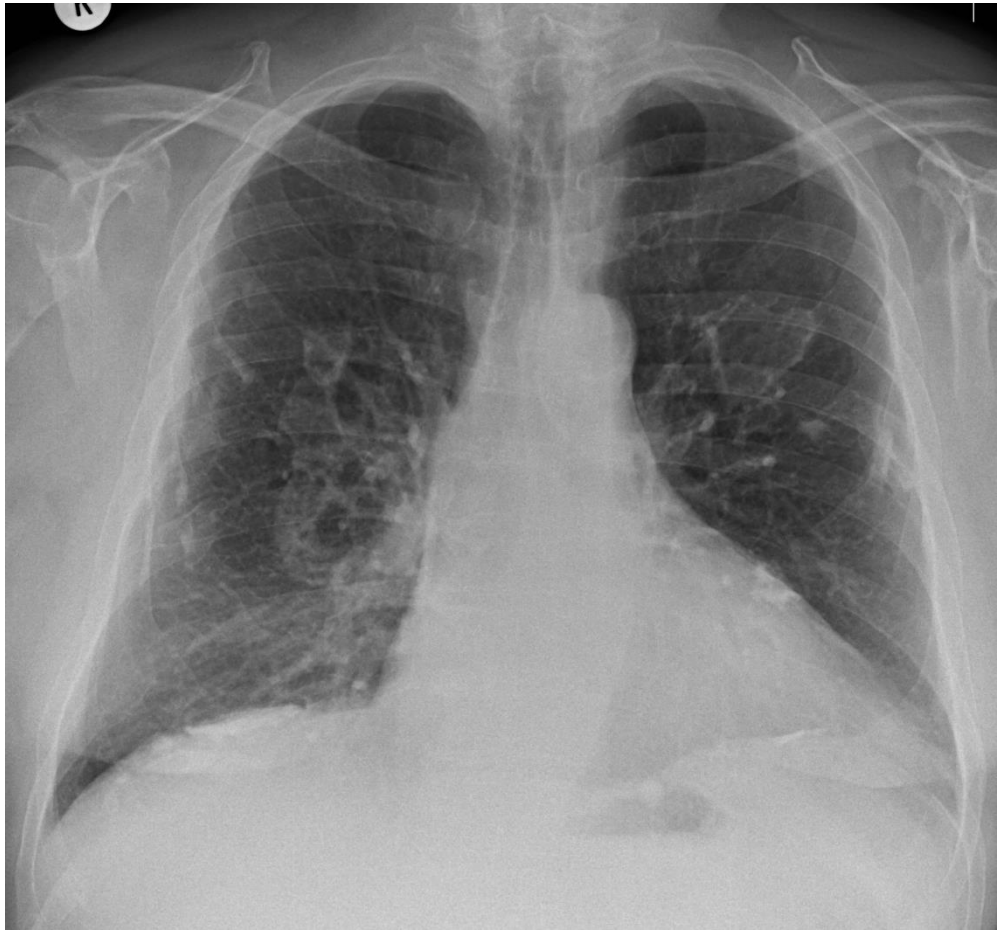
An ILD doctor

69M
smoker
cough
CXR

- site carpenter: 16-65

- (OA knee
- hypertension)



What will you tell him?

- a plaque is a ‘memory’ of your work with asbestos …
- … and no more than that

- (you don’t have ***asbestosis)***

PH: CXR report

1377632 12/08/2014 Chest XR

There is bilateral pleural calcifications suggesting previous asbestosis exposure.
The heart is slightly enlarged.
No active lung lesions are seen.
The pleural spaces are clear

What will you tell him?

- a plaque is a ‘memory’ of your work with asbestos …
- … and no more than that

- (you don’t have ***asbestosis)***

- the plaques won’t grow …
- … won’t cause you any symptoms …
- … and won’t turn into cancer
- ….same risk of mesothelioma as an identical carpenter without plaques

- (but … a man of his age in the UK has 1% chance of mesothelioma)

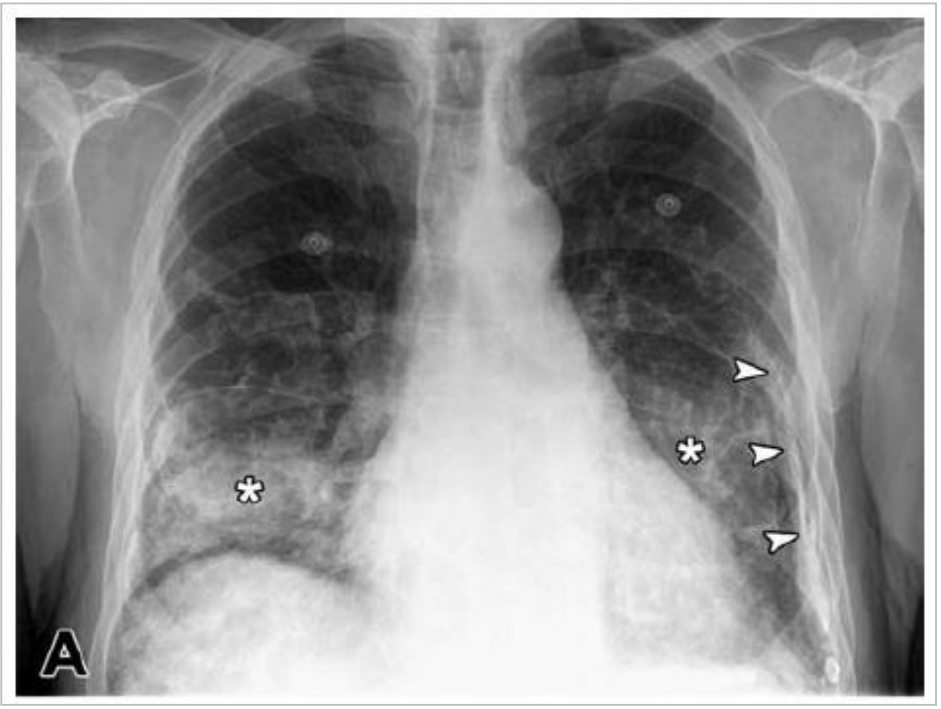
- compensation …

When is IPF asbestosis?

When is IPF asbestosis?

- asbestos exposure was very common in UK men
- (still common outside Europe)
- all types of asbestos give rise to asbestosis …
- … at high doses: **exposure history**
- CT is highly sensitive
- the pathology is not specific (UIP)
- look for accompanying radiological evidence of asbestos exposure
- some decisions can be very hard
- ‘the balance of probabilities’

‘easy’ case



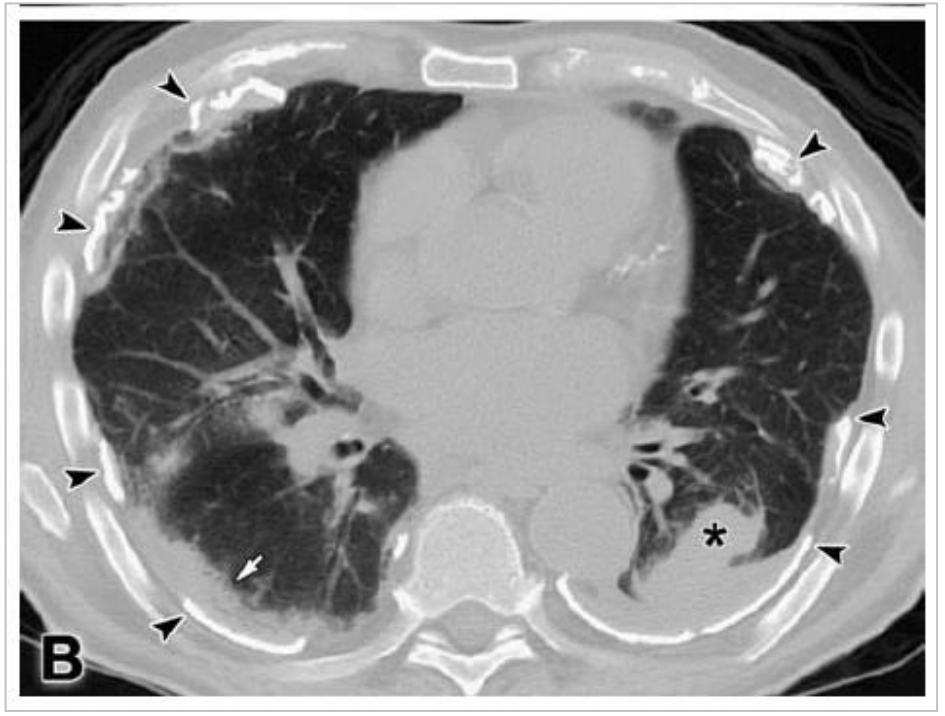


Table 2. Differential Diagnostic Features for Asbestosis and Idiopathic Interstitial Pneumonias

Histologic Feature	UIP	Asbestosis	NSIP
Distribution	Subpleural accentuation, lower lung zone	Peribronchiolar with subpleural accentuation	Diffuse
Honeycomb changes	Common	Uncommon except in advanced cases	Uncommon
Fibroblast foci	Conspicuous	Rare	Inconspicuous
Asbestos bodies	Absent	Frequent[a]	Absent
Inflammation	Minimal, typically localized to honeycomb foci	Minimal	Variable
Pleural fibrosis[b]	Uncommon	Common	Uncommon

10%

Asbestos fibre concentrations in different industries (Germany)

Work area		1950–54[a]	1970–74	1980	1990
Textile industries	FRG	100	10	3.8	0.9
	GDR	100	12	6.2	2.2
Production of gaskets	FRG	60	6.6	4.7	0.7
	GDR	60	8.0	7.8	1.6
Production of cement	FRG	200	11	1.1	0.3
	GDR	200	13	1.9	0.7
Production of brake pads	FRG	150	9.1	1.4	0.7
	GDR	150	11	2.4	1.6
Insulation works	FRG	15	15	8.6	0.2
	GDR	18	18	14.0	0.5

‘Helsinki criteria’: 25 f/ml-years

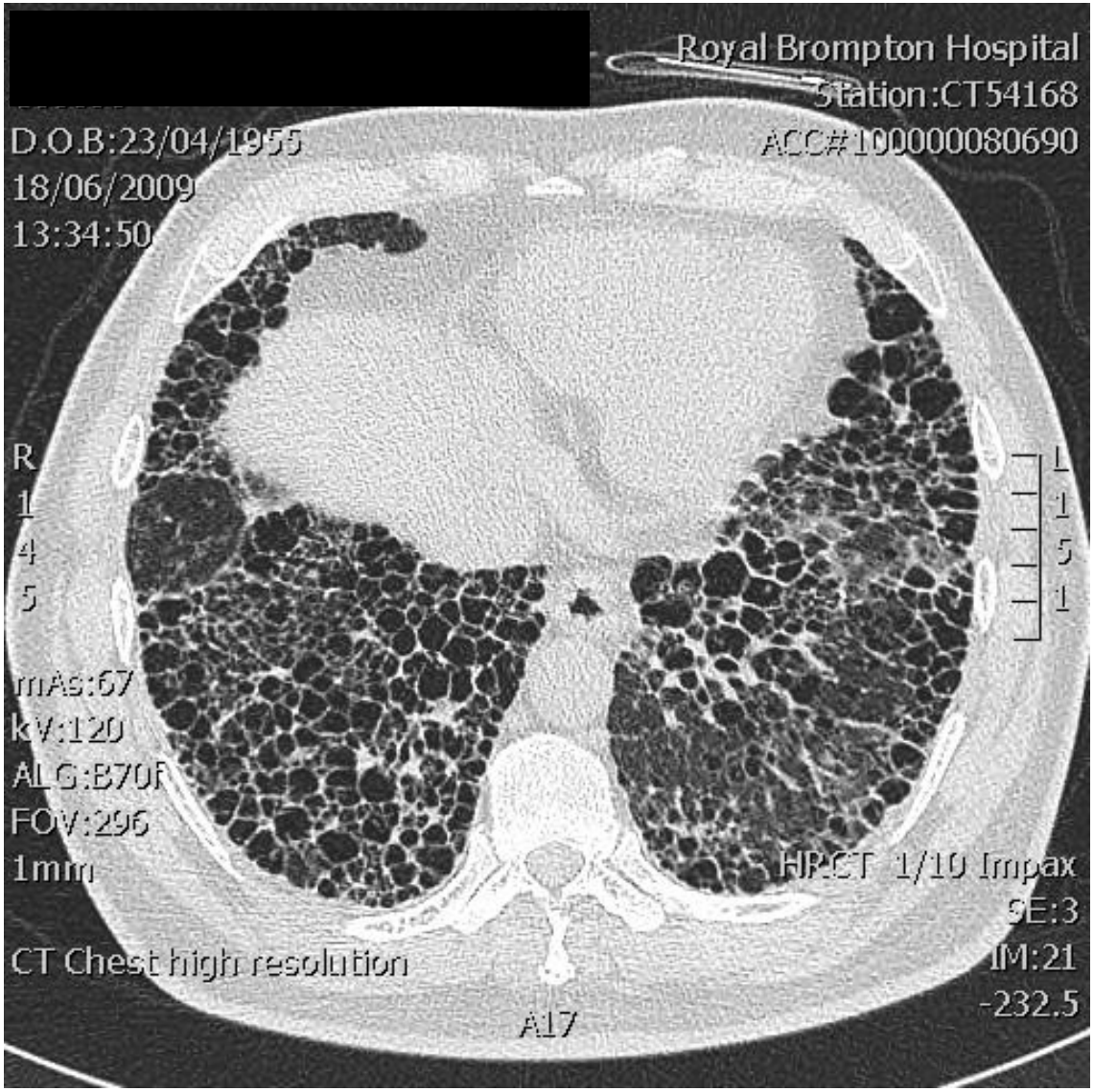
German Democratic Republic (E)
Federal Republic of Germany (W)

One for you to try

- man of 57

- ‘IPF’: CT and biopsy

- 1983/4 Bingley Son and Follett (Park Royal)
- cutting and fashioning asbestos sheeting
- dusty++
- casual work at same place for 2 years further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CT Chest high resolution
A17

Taking a history

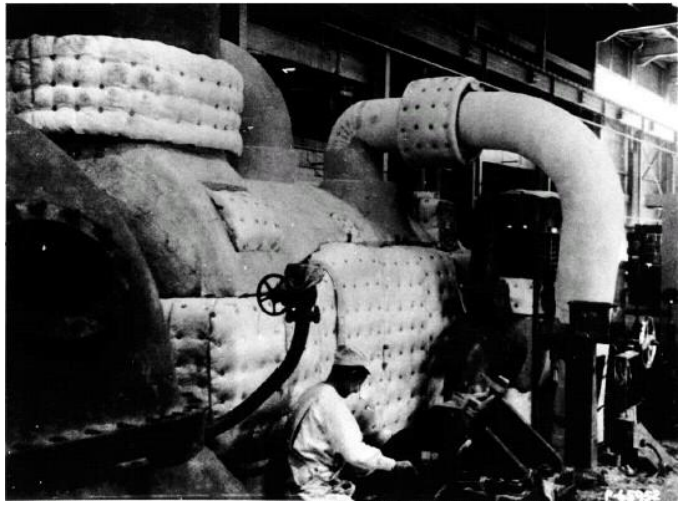
Last year, my neighbour's father died from mesothelioma

- he was a photographer

Taking a history

Last year, my neighbour's father died from mesothelioma

- he was a photographer: for the Central Electricity Generating Board



Newcastle shipyard in 1940s

80M Lagger

1950 to mid 1980s worked in East End
Minimal smoking history

15 years of SOBOE and occasional productive cough

Wife recently told she has pleural plaques

Crackles on auscultation

CXR pleural plaques

FEV1 2.64 (88%)
FVC 3.54 (88%)
FEF 25-75 60%

What is the diagnosis and what would you do next?

Diagnoses:
1. Asbestos related pleural plaques
2. Probable COPD

Recommended medication:
Ultibro one daily.

I saw this gentleman in the clinic today. He is a gentleman with previous asbestos exposure with known asbestos related pleural plaques. I understand that his wife has recently been diagnosed with pleural plaques which is probably occupational.

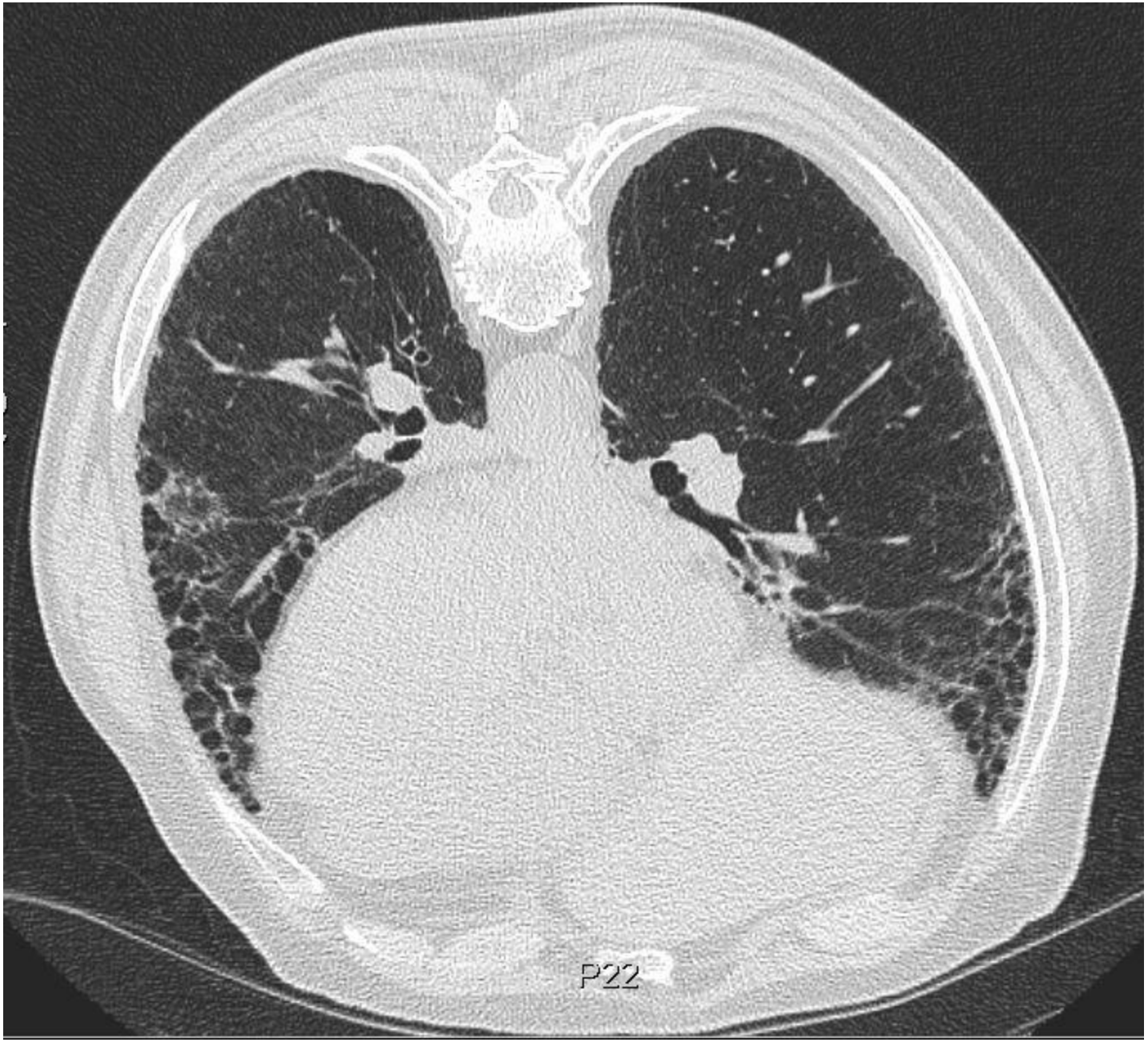
He has noticed that over the last year his exercise tolerance has decreased by about 50% and he does get breathless on climbing stairs (MRC breathless score 3).

Spirometry today in the clinic shows his basic FEV1 and FVC to be well preserved with an FEV1 of 2.86 L and FVC 3.06 L, but on examining him he has a prolonged expiratory phase and his FEF 25-75 was 60% predicted suggesting a degree of small airways disease.

He stopped smoking at the age of 45 and I therefore think that there may be a COPD element to this. I would therefore suggest that he is started on LAMA/LABA and you give him at least three months trial of this.

He has indicated that he was wondering whether he can consider applying for asbestos related disease. I have advised him that following an appeal court ruling that there were no grounds for civil or statutory compensation for asbestos related pleural plaques.

Clinically I do not think that he has any fibrosis (asbestosis), but if you are concerned about this then I would suggest that you consider high resolution CT scan locally.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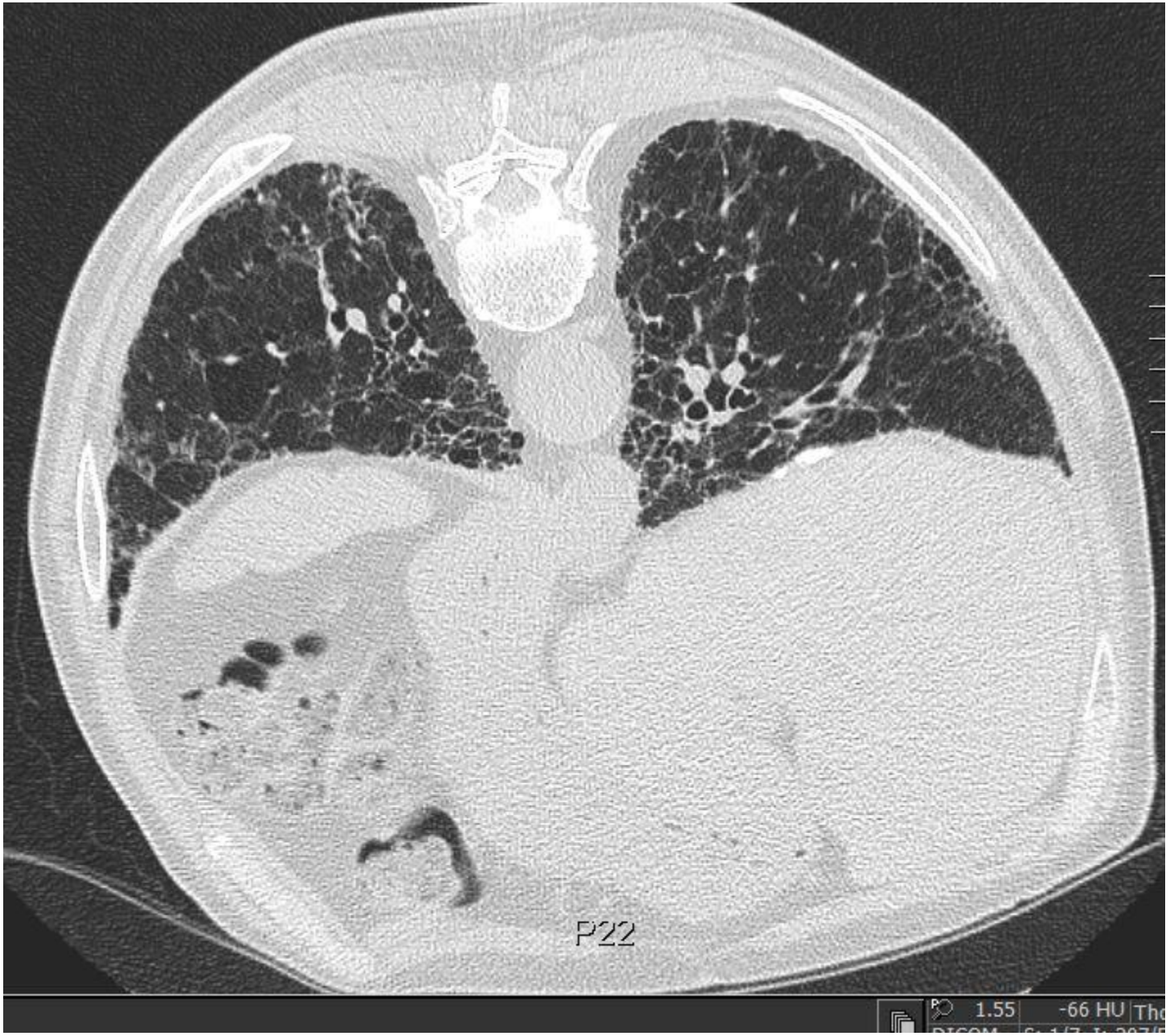
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1.55
-66 HU

Radiology report

There is extensive coarse reticular shadowing most marked in the lower lobes but extending also into the upper lobes. There are calcified pleural plaques over the left hemidiaphragm. No other abnormality noted.

The parenchymal changes are more suggestive of IPF rather than asbestos-related interstitial lung disease although I cannot exclude this.

Our view

- Asbestosis (not IPF or COPD)
- Advised eligible to apply for IIDB
- Eligible to make a claim for legal compensation (Union)

What causes silicosis?

A. silicon

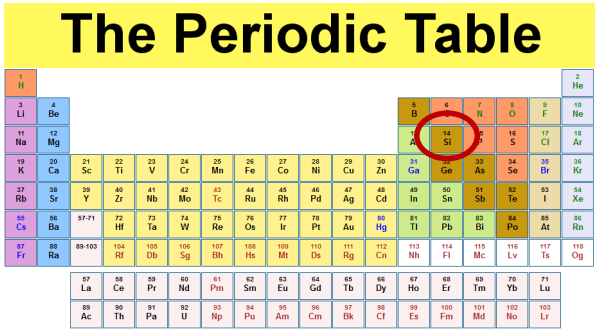
B. silica

C. silicone

D. rcs

What causes silicosis?

A. silicon = Si, the element



B. silica

C. silicone

D. rcs

What causes silicosis?

A. silicon = Si, the element

B. silica – silicon dioxide (SiO_2 - ‘quartz’)

C. silicone

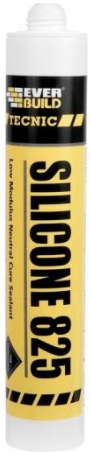
D. rcs

What causes silicosis?

A. silicon = Si, the element

B. silica – silicon dioxide (SiO_2 - 'quartz')

C. silicone = polymer of O_2 and Si



D. rcs

What causes silicosis?

A. silicon = Si, the element

B. silica – silicon dioxide (SiO_2 - 'quartz')

C. silicone = polymer of O_2 and Si

D. rcs = *respirable crystalline silica*

Silicosis

Adverse pulmonary response to high, accumulated doses of inhaled RCS

Long latency disease (most after 15 years)

Approximate crystalline silica content of different materials	
Sandstone	70–90%
Concrete, mortar	25–70%
Tile	30–45%
Granite	20–45%, typically 30%
Slate	20–40%
Brick	Up to 30%
Limestone	2%
Marble	2%

Why don’t you get silicosis lying on the beach?

HORSE Journals

J Vet Intern Med. 1991 Jul-Aug;5(4):248-56.

Thoracic radiographic features of silicosis in 19 horses.

Berry CR[1], O'Brien TR, Madigan JE, Hager DA.

Author information

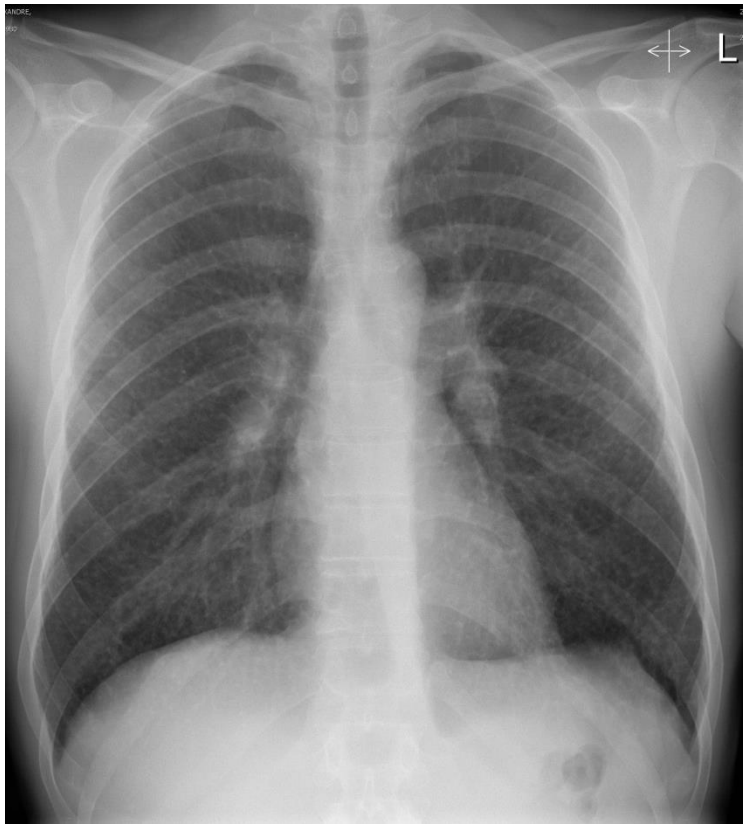
Abstract

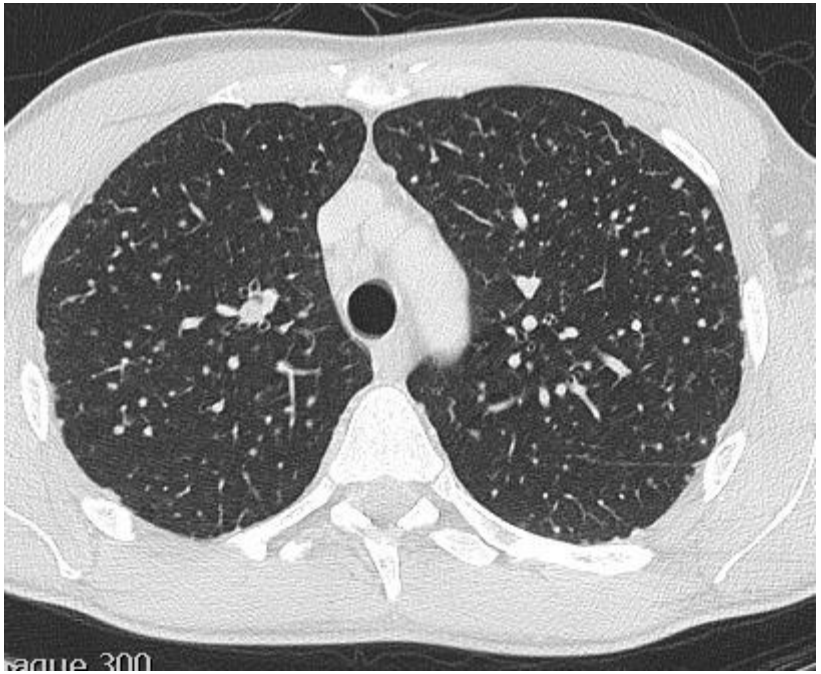
Clinical records and thoracic radiographs of 19 horses with a confirmed pathologic diagnosis of silicosis were reviewed. These horses had histories of varying degrees of chronic weight loss, exercise intolerance, and respiratory distress. At the time of presentation, two horses were asymptomatic. Ten horses were geldings and nine were female. The mean age of the 19 horses was 10.7 +/- 5.5 years. Fourteen horses were identified as being from the Monterey-Carmel Peninsula of midcoastal California. An abnormal, structured interstitial pulmonary pattern was identified on thoracic radiographs in each horse. The interstitial pulmonary changes were classified as miliary (13 horses), reticulonodular (4), or linear interstitial (2), and were best visualized dorsally and caudodorsally. In addition to the abnormal interstitial pulmonary pattern, areas of pulmonary consolidation were evident caudodorsally in seven horses. Other thoracic radiographic features included: hilar lymphadenopathy (4 horses), pleural effusion/thickening (4), cranial mediastinal lymphadenopathy (2), hyperinflation (1), and a discrete pulmonary mass (1). Necropsy findings in eight horses and results of lung biopsies in an additional five horses showed a diffuse, multifocal, granulomatous pneumonia with areas of pulmonary fibrosis. Cellular infiltrates included predominantly macrophages with intracellular and/or extracellular crystalline material, occasional lymphocytes, and giant cells. Similar cellular changes were also identified, during necropsy, in the hilar and tracheobronchial lymph nodes in each of the eight horses, although gross enlargement of the lymph nodes was present in only six horses. The radiographic and pathologic findings of these 19 horses are consistent with chronic or the accelerated forms of silicosis that are recognized in humans.

PMID: 1941758

[Indexed for MEDLINE] **Free full text**

Simple silicosis vs complicated (PMF) silicosis

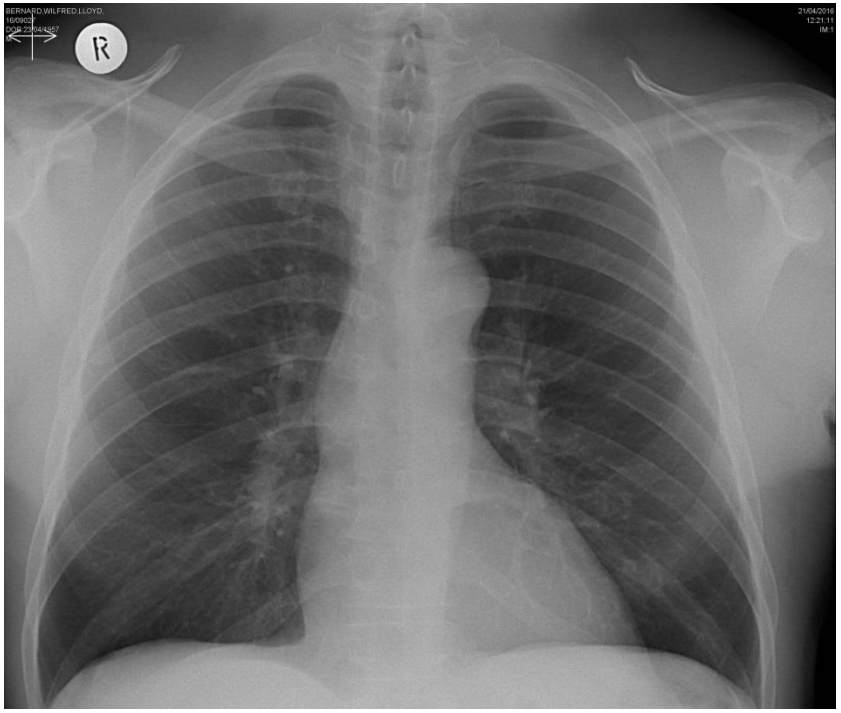




Portugese stonemason
asymptomatic & normal lung function

59 year old train driver

- no respiratory symptoms
- job entails filling a sandbox under his locomotive



Clinical details: History of exposure to silica.

“There are multiple small dense nodules in the mid and lower perihilar regions which may represent calcified pulmonary nodules. Linear atelectasis in the right mid zone but otherwise no other lung change.
The above described findings may represent previous exposure to silica.”

Reported By: Dr X Consultant Radiology

spirometry c.35% predicted …..

55M Stonemason

Poland, Germany, UK for 10 years

Works uniquely with limestone now

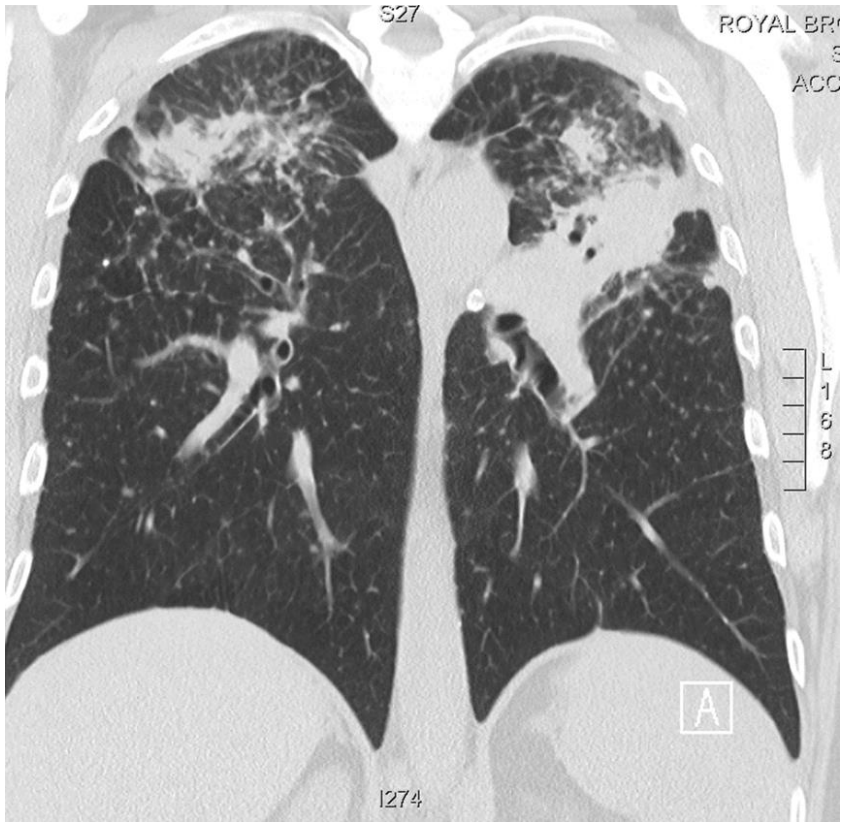
Incidental finding of abnormal CXR on routine surveillance

No respiratory symptoms

Occasional night sweats; unquantifiable weight loss

No medical history of note; no drugs; never smoked

What is the diagnosis and what to do you need to do to confirm it?



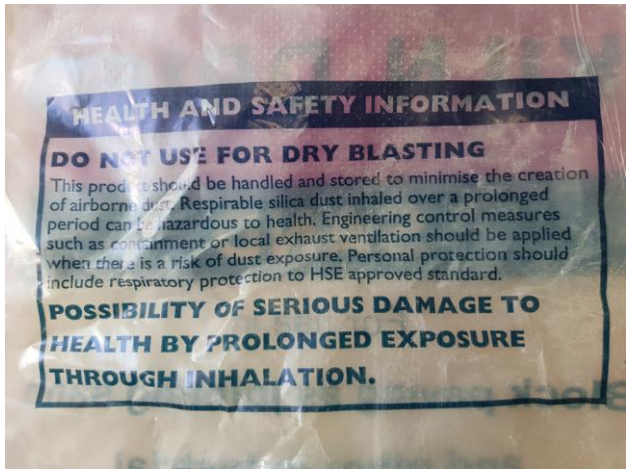
65M works in motor vehicle industry

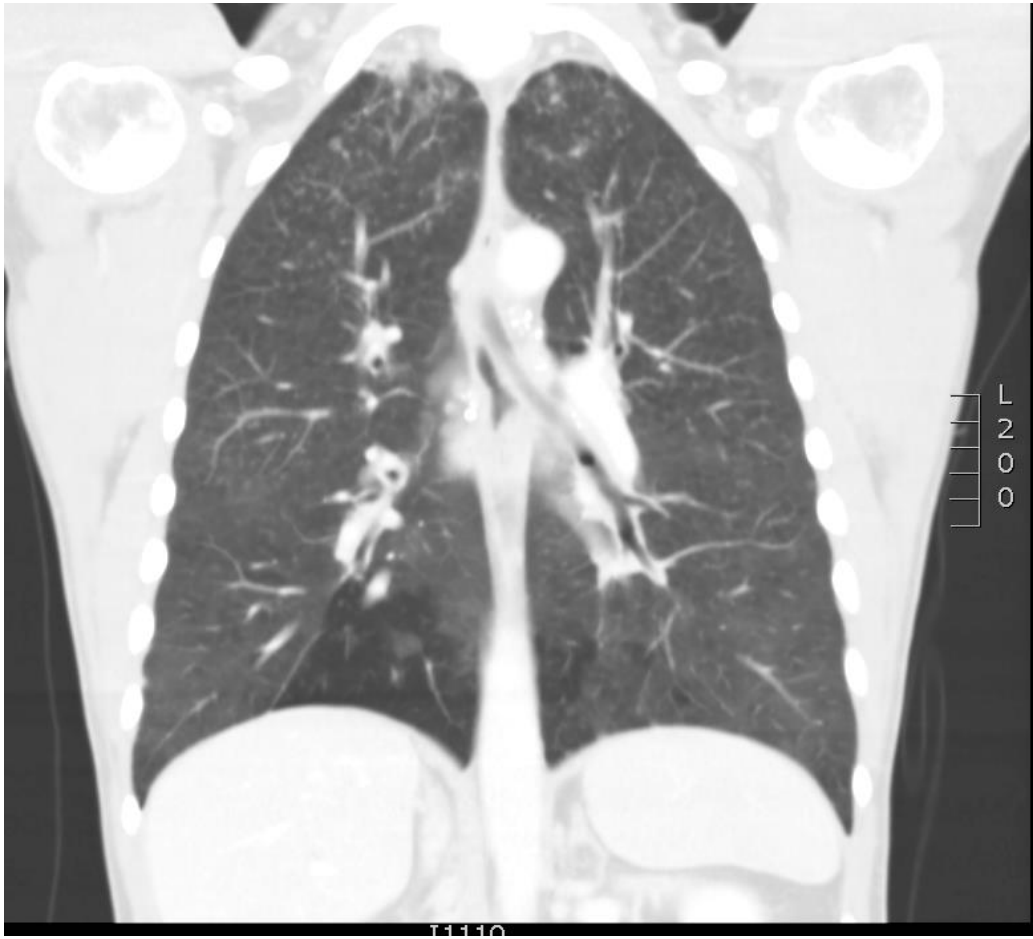
sandblasts vintage cars

in same job for 37 years

uses this sand -

moreover, it is recycled…..





What would you advise?

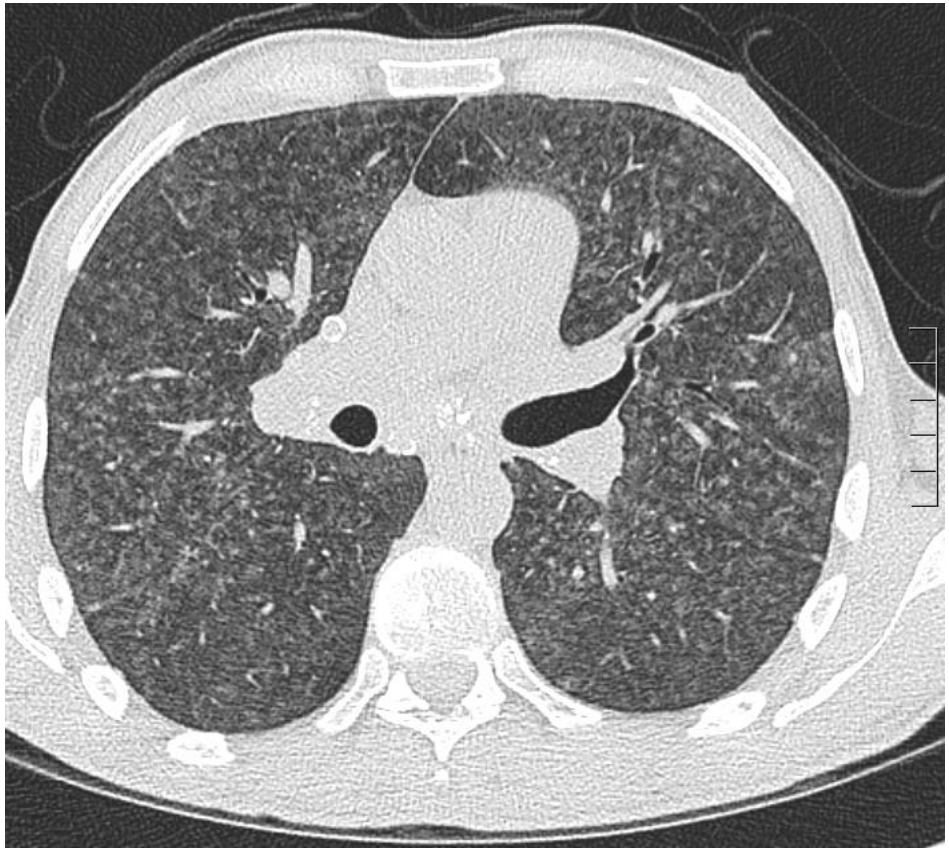
LEV installed and RPE worn

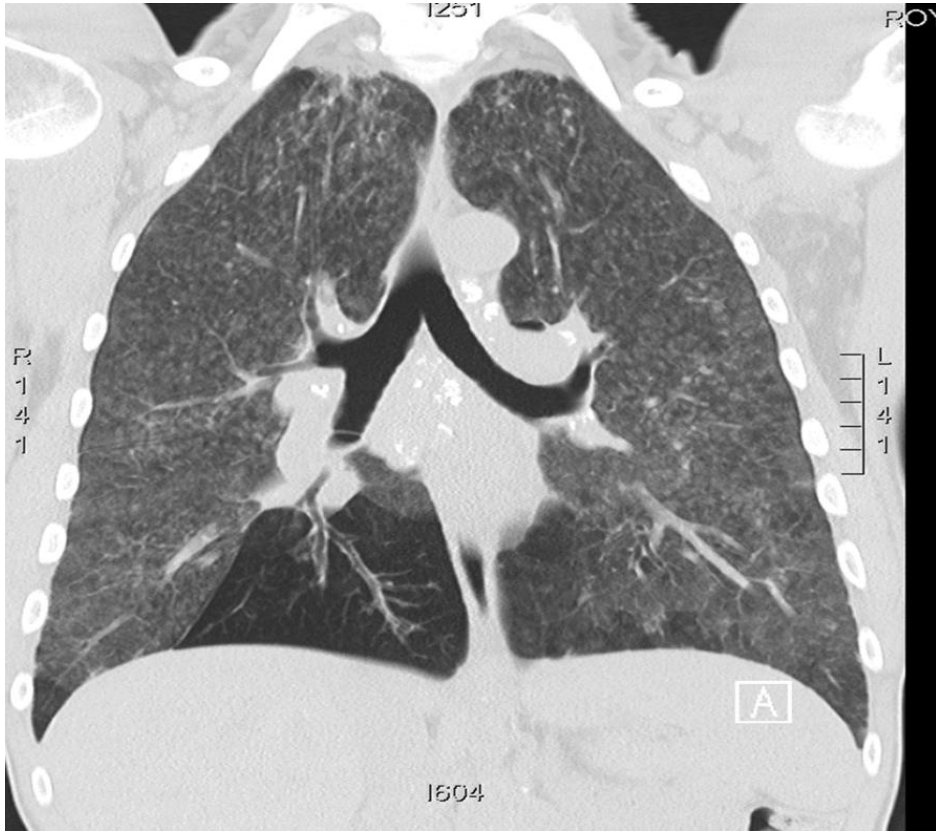
One year later, re-presented locally with worsening symptoms
Referred to OccLD clinc

Worsening dyspnoea, weight loss, night sweats

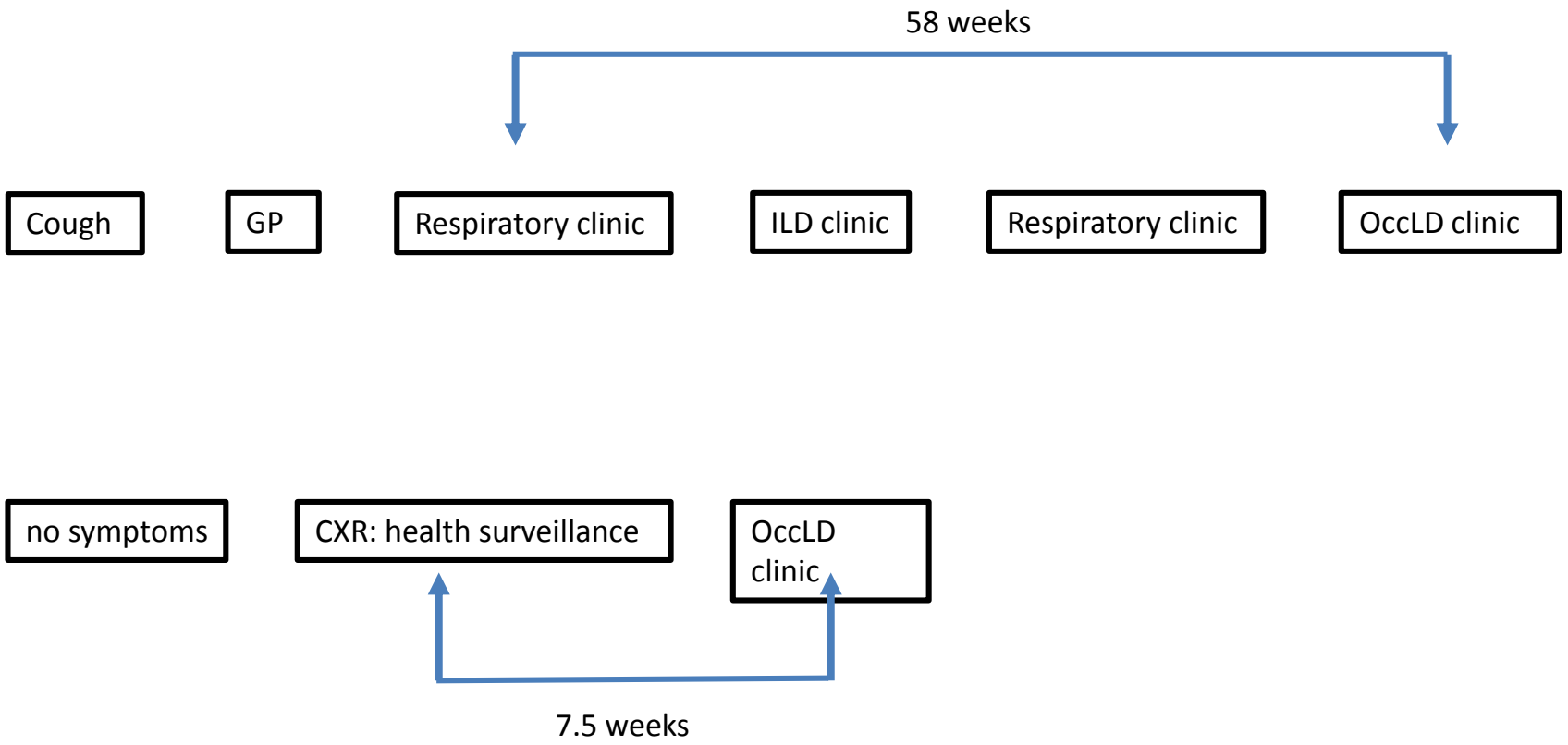
Differential diagnosis?

What would you advise now?





Contrasting pathways



What (fairly) common pulmonary disease looks like CBD?

CBD = chronic beryllium disease

When is sarcoidosis actually chronic beryllium disease?

- 1934 zinc-beryllium silicate phosphor for more light output with better white colours
- 1946 Harriet Hardy: ‘Salem sarcoid’ n=17 in a lightbulb factory (3 ‘community’)

CBD as ‘sarcoidosis’?

- it’s all pulmonary
- isolated BHL does not seem to occur
- they have a positive BeLPT (fresh blood or BAL)

  - measures sensitisation to Be
  - highly specific
  - don’t need much exposure
  - most who are sensitised will get CBD
  - surveillance in US but exposure minimal in the UK

How often is sarcoidosis actually CBD?

	Israel	Germany/Israel
	Fireman SVDLD 2003	Müller-Quernheim ERJ 2006
# sarcoid	47	536
# 'exposed'	14 (30%)	84 (16%)
# BeLPT	14	84
CBD	**3 (6%)**	**34 (6%)**

exposed industries (Germany/Israel)

Individuals	34
Dental technician/dentist	13 (7/6)
Engine development/mechanics/ automobile industry	2 (1/1)
Brass alloys, beryllium-containing alloys[#]	4 (4/0)
Metallurgic factory	2 (1/1)
Aircraft production and maintenance	3 (2/1)
Nonsparking tools	1 (1/0)
Radiation shielding	1 (0/1)
Military vehicle armour	2 (1/1)
Fluorescent lamps	2 (1/1)
Microelectronics/electrical relays	1 (1/0)
Chemical industry[¶]	1 (1/0)
Engraving of gems	1 (1/0)
Ore mining	1 (1/0)
Grinding of optical lenses for precision instruments	

Müller-Quernheim ERJ 2006

How often is sarcoidosis actually CBD?

	Israel	Germany/Israel	**Ontario**
	Fireman SVDLD 2003	Müller-Quernheim ERJ 2006	Ribeiro Lung 2011
# sarcoid	47	536	121
# 'exposed'	14 (30%)	84 (16%)	34 (28%)
# BeLPT	14	84	34
CBD	**3 (6%)**	**34 (6%)**	**0**

Consider

Referral from A Respiratory Consultant

42F works in Sainsbury's bakery department

Started job last autumn
Developed cough and SOB on exposure to flour; no mask
Symptom free when not at work
Has not seen OH
Smoker 10cpd

Best PEFR 380; previously as low as 210 at work

Started her on Ventolin and Serevent
Suggested that she moves away from the bakery

The patient recalls …

- she told me I had baker's asthma …
- … and should leave my job

What would you have done?

1. a careful history

In 2015 she started to work in the X branch of Sainsburys, initially on the checkouts, but in September 2016 she moved to the bakery. Initially, she was based in the confectionary area which involved cake decoration but no direct contact with flour; four months ago she started to work as a baker. Following her recent appointment with you she has moved temporarily back into the confectionary area.

2. Is she sensitised?

Skin prick tests were positive to extracts of grass pollen (4 mm) and negative to extracts of cat, house dust mite, wheat flour, plain flour, alpha amylase, soy and rice flour antigen with appropriate positive and negative controls. Serum specific IgE to plain flour and alpha amylase were negative

3. Does she have asthma?

Her spirometry was within normal limits: FEV_1 2.38 (101%), FVC 3.01 (109%) with no significant changes following bronchodilator therapy

What we did next

- Her immunology tests confirm that she is not sensitised to bakery allergens and that I think it is *highly unlikely* that her symptoms are due to baker's rhinitis or asthma.

- It is not uncommon for bakers to report nasal symptoms but this is often due to irritation from flour dust rather than a specific sensitisation.

- I have not seen any evidence to suggest that she has a diagnosis of asthma although I note she reports subjective improvement with Serevent.

- I have suggested that she *stops her inhalers and that she returns to work in the bakery*. She is going to complete detailed peak flow monitoring every two hours on days at work and days away from work for four weeks which should allow us to better understand what is causing her symptoms.

- I will review her again in six weeks.

- Update: no symptoms since stopping inhalers; not allowed to return to bakery……..

44 FT firefighter

Fit and well
Grenfell Tower June 2017
Acute bronchitis symptoms for 2 weeks after

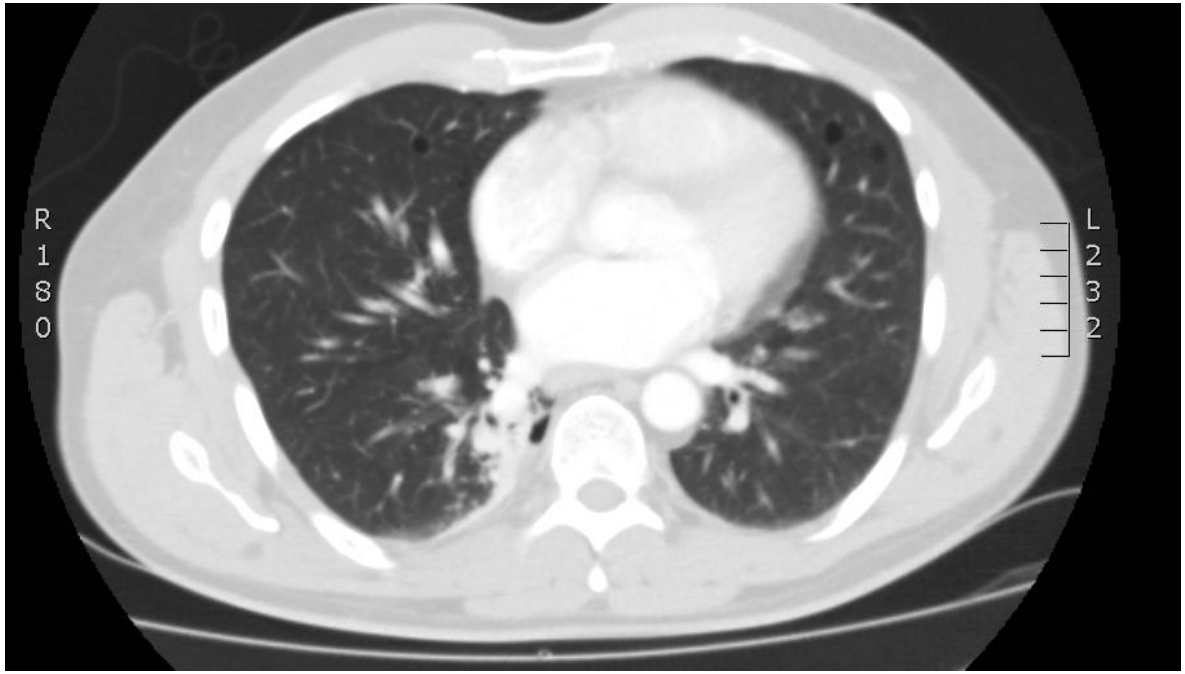
Progressive exertional dyspnoea over the autumn
Couldn't do fitness tests

October 2017
Develops acute SOB and diagnosed with pneumothorax
Failed aspirated, wide bore tube.
Discharged

Light duties
Fitness slowly returns

Referred to OccLD clinic: implications of a pneumothorax for a FF

What would you do next?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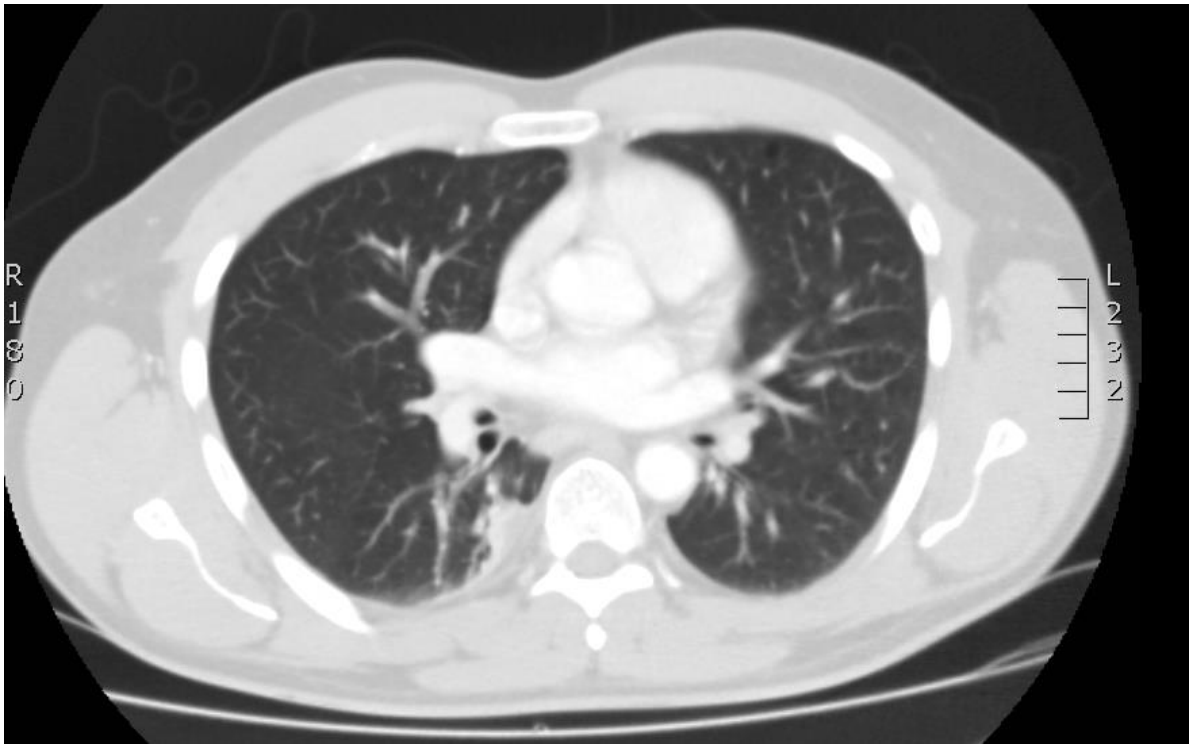
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Referred to OccLD clinic: implications of a pneumothorax for a FF

What would you do next?

Review of CT

Small number of thin-walled cysts

Differential diagnosis?
LIP (CTD antibody profile)
HIV
Birt Hogg Daub syndrome

Heterozygous for a known pathogenic frameshift variant in the *FLCN* gene: c.1285delC, p.(His429ThrfsTer39), which is predicted to cause loss of function via premature protein truncation, a known disease mechanism for *FLCN*.

This variant is reported as one of the commonest pathogenic variant causing BHD. It lies in a known mutation hotspot, and has been reported in multiple BHD patients (OMIM allelic variant 607273.0001). Nickerson *et al* (*Cancer Cell* 2: 157-164, 2002) detected this variant in 29% of patients with clinically diagnosed Birt-Hogg-Dube patients.

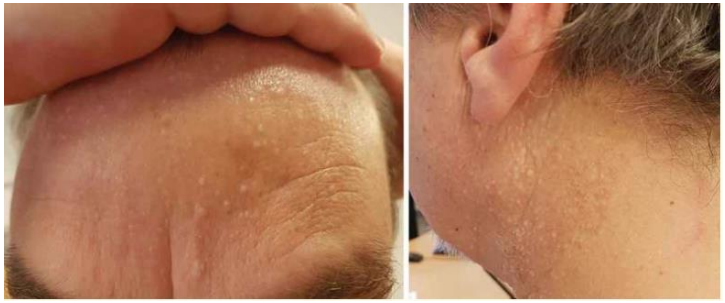
The variant has also been reported as pathogenic by multiple clinical laboratories (ClinVar variation ID 3363). The variant has also been reported in the gnomAD control population (9/108360 alleles in the gnomAD European population).

This result confirms a clinical diagnosis of BHD, and appropriate clinical surveillance is recommended.

BHD syndrome

Autosomal dominant disorder caused by mutations in
the *FLCN* gene which is a tumor suppressor gene and codes for the protein folliculin

Prevalence unknown (n=600 families described)



Associated with fibrofolliculomas (58-90%)
Renal cell carcinoma (30%; x7fold increase)
Cystic lung disease (67-90%; 40-75% PTx)

- n=0 to 407
- bilateral, basal, irregular shaped, thin walls

Family screening
Annual renal MRI
No way of predicting if further pneumothoraces
Refer for surgical opinion (nb no progressive respiratory failure)

Other issues:
Unwilling for us to liaise with OH
? fit to continue to work

Another case

May 2011

Dear Paul

Please would you see this baker ....

1st June 2011
(Clinic – 25.05.11)

Occupational Health Manager
Head Office

Dear Sir/Madam

Re:

Diagnosis: Extrinsic allergic alveolitis – precipitating factor working in a bakery

This 51 year old lady works in the Harpenden branch of within the bakery department. Prior to joining she had been completely well. After starting work in the bakery nearly a year ago she noticed that during the day she became progressively breathless and felt run down with some nasal stuffiness. She then had some attacks of sudden onset breathlessness during her work in the bakery which ended up her having to go home early from work.

She first attended the chest clinic in March 2011 and we have been investigating the cause of this breathlessness and I am sorry to say that this appears to be a case of extrinsic allergic alveolitis. This is a condition where the body reacts with an abnormal allergic reaction to a certain precipitant causing the symptoms that Mrs has been describing. In this case it would appear that something in the bakery, be it one of the grains or indeed the yeast is causing her to have these horrible allergic reactions. Unfortunately the only treatment for this condition is complete avoidance of the precipitant. She has brought to me today a peak flow diary which shows significant deterioration in her breathing whilst in the working environment and I think this confirms the diagnosis.

I would be extremely grateful if you could arrange for this lady to be moved out of the bakery and for her future health it is important that she never works in the bakery again. This will completely cure her of the condition. If she were to continue working in the bakery she will develop permanent lung disease.

Many thanks for your consideration and please do not hesitate to get in touch with me if you require further information.

Yours sincerely

Another case

- is this EAA?.
- is EAA a recognised disease among bakers?
- if it isn’t EAA what is it?
- what next?

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The end