ANZSRS Case of the Month - January 2024

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Pulmonary Silicosis caused by Engineered Stone

Background

Silicosis is an occupational lung disease that has been seen as far back as 430 B.C by Hippocrates, in 16th century by Agricol and in 1713 by Rammazini who found silicotic nodules in post-mortems of stone cutters[1]. Historically, it has been a lung disease that has affected people who work with silica in its natural environment such as miners, tunnels, and quarry workers. In developed countries, there has been a decline in silicosis-related mortality in these sectors due to significant changes in occupational health and safety measures[1]. Documentation of cases of pulmonary silicosis caused by engineered stone (ES) began world-wide in 2009[1] and cases have been on the rise in Australia since 2015[2]. ES, used in benchtop fabrication, is made of finely crushed rock, with a crystalline silica content > 90%, pigments and polymeric resins acting as binders[2,3]. Through the process of cutting and grinding the stone, it exposes workers to fine particles which accumulate in the lungs, causing inflammation, cytotoxicity and ultimately leading to silicosis; a type of pulmonary fibrosis[4]. When compared to lung disease caused by natural silica, ES silicosis has a short latency and rapid progression from simple to progressive massive fibrosis and has higher mortality rates[2.3]. Simple silicosis is characterised by nodules <1cm in size, usually predominant in the upper lobes. Common symptoms include exertional dyspnoea, chronic cough, and sputum production. Simple silicosis can progress to progressive massive pulmonary fibrosis (PMF), which is characterised by silicotic nodules >1cm, other possible progressions include central cavitation, enlarged hilar or mediastinal lymphadenopathy, significant distortion of surrounding lung parenchyma and peribronchial vessels and pleural thickening. A large retrospective study found that the progression from simple to PMF occurs in 18-37% of silica workers over an average of 5 years[1].

In a recent study in Australia, of 544 workers screened, 117 (28.3%) were diagnosed with silicosis (96 simple and 21 complicated), all male and more than half had migrated to Australia, with a median age of 42.1 years and an average exposure time of 12 years[3]. In addition, silicosis has been linked to increased risk of autoimmune diseases such as, rheumatoid arthritis, systemic sclerosis, psoriatic arthritis, abnormal autoimmune serology, Sjogren's syndrome, antineutrophil cytoplasmic antibody associated vasculitis, systemic lupus erythematosus and mixed connective tissue disease[5].

Since 2015 there have been 570 cases of silicosis in Australia linked to ES. Due to the high risk of disease to those working with the material, on the 13th of December 2023, the Australian Government unanimously agreed to completely ban the use, supply, and manufacturing of the product. This was a world-first decision, with the ban going into effect from 1st of July 2024[6,7].

Case Presentation

Here we present a 41-year-old man, originally from Indonesia who migrated to Melbourne, Australia in 2008 where he was working with engineered stone from 2008 – 2020. Ten of those years he was dry cutting stone with paper masks. He was diagnosed with simple silicosis in 2020 and transferred care from Victoria to Adelaide in 2021. Other problems included a history of Tuberculosis lymphadenitis in 2017, likely related to silicosis, which was treated for 12 months.

2020 (Victoria)

Lung Function

FEV_1 post-BD 3.24 L (91%), FVC 3.82L (94%)

D_LCO Hb-corrected 23.8 ml/min/mmHg(80%), KCO Hb-corrected 4.43 min/mmHg (95)

HRCT

Numerable nodules present within both the upper and lower lobes of the lungs with slightly upper lobe predominance, no features to suggest underlying progressive massive fibrosis. Overall findings suggestive of silicosis

2021 – 2022

December 2021

Symptoms: Patient struggled with coughing every morning and production of clear sputum. Shortness of breath with strenuous exercise

February 2022

HRCT

Nodules unchanged in size, number, and configuration from earlier study

August 2022

Symptoms: Increased tiredness and increased shortness of breath, noticed during gardening e.g., mowing the lawn

Lung Function

Spirometry stable from previous testing in 2020, DLCO stable in 2021 and declined in 2022.

SEQUENTIAL DATA

Date	Pre BD FEV_1 (L)	FVC (L)	FEF_{25-75} (L/s)	Post BD FEV_1 (L)	FVC (L)	FEF_{25-75} (L/s)
09-12-2021 R	2.86 (78%)	3.47 (76%)	2.85 (79%)	3.05 (83%)	3.75 (83%)	2.97 (82%)
24-08-2022 R	3.00 (83%)	3.76 (84%)	2.76 (78%)	3.08 (85%)	3.76 (84%)	3.29 (92%)

Date	Uncorrected D_LCO (mL/min/mmHg)	Corrected D_LCO (mL/min/mmHg)	Corrected KCO (mL/min/mmHg)
09-12-2021 R	22.1 (80%)	21.9 (80%)	4.3 (92%)
24-08-2022 R	19.6 (72%)	19.2 (71%)	3.9 (82%)

Date	VC_{Pleth} (L)	RV_{Pleth} (L)	TLC_{Pleth} (L)	RV/TLC_{Pleth}	FRC_{Pleth} (L)
09-12-2021 R	3.95 (81%)	1.43 (98%)	5.37 (85%)	27	2.69 (94%)
24-08-2022 R	3.86 (80%)	1.52 (105%)	5.35 (86%)	29	2.86 (101%)

2023

February

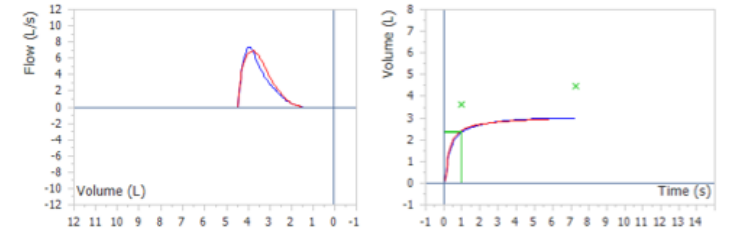
Patient reviewed with repeat lung function testing and HRCT.
New symptoms: chest tightness for two weeks. Lung function and HRCT confirm simple silicosis to progressive massive silicosis.

Lung Function

Significant decline in all parameters

Forced Expiratory Volumes (BTPS) [09:16] GLI2012

		Pre BD		Post BD		%Change	Predicted Mean (Range)	Z-Score
FEV_1	(L)	2.36	(65%)*	2.44	(68%)*	4%	3.62 (>2.86)	-2.50
FVC	(L)	2.95	(66%)*	2.90	(65%)*	-2%	4.48 (>3.55)	-2.80
FEV_1 /FVC		80%		84%			81% (>70%)	0.59
FEF_{25-75}	(L/s)	2.20		2.88		31%	3.55 (>2.02)	-0.67
FEF_{50} /FIF_{50}		65%		97%				



SUBDIVISIONS OF LUNG VOLUMES (Plethysmography) [09:44] GLI2021

		Observed		Predicted Mean (Range)	Z-Score
VC	(L)	2.98	(62%)*	4.82 (3.86-5.80)	-3.20
IC	(L)	2.06	(61%)*	3.40 (2.42-4.34)	-2.23
ERV	(L)	0.88	(70%)*	1.26 (0.55-2.21)	-0.81
RV	(L)	1.45	(99%)*	1.47 (0.83-2.25)	-0.05
TLC	(L)	4.39	(70%)*	6.28 (5.08-7.49)	-2.60
RV/TLC		33%		23% (14%-33%)	1.72
FRC	(L)	2.33	(81%)*	2.86 (1.95-4.01)	-0.92

SINGLE BREATH DIFFUSING CAPACITY [09:19] GLI2017

		Observed		Predicted Mean (Range)	Z-Score
$D_LCO_{uncorrected}$	(mL/min/mmHg)	17.1	(63%)*	27.09 (20.97-34.20)	-2.83
$D_LCO_{corrected\ for\ Hb}$	(mL/min/mmHg)	17.0	(63%)*	27.09 (20.97-34.20)	-2.86
$KCO_{corrected\ for\ Hb}$	(min/mmHg)	4.9	(103%)*	4.72 (3.70-5.83)	0.23
V_A	(L)	3.5	(60%)*	5.78 (4.71-6.92)	-3.71
IVC	(L)	2.8	(62%)*	4.48 (3.55-5.43)	-2.98

HAEMOGLOBIN

		Observed	(Range)
Hb	(g/L)	148	(135-175)

SEQUENTIAL DATA

Date	Pre BD FEV_1 (L)		FVC (L)		FEF_{25-75} (L/s)		Post BD FEV_1 (L)		FVC (L)		FEF_{25-75} (L/s)	
09-12-2021 R	2.86	(78%)	3.47	(76%)	2.85	(79%)	3.05	(83%)	3.75	(83%)	2.97	(82%)
24-08-2022 R	3.00	(83%)	3.76	(84%)	2.76	(78%)	3.08	(85%)	3.76	(84%)	3.29	(92%)
14-02-2023 R	2.36	(65%)	2.95	(66%)	2.20	(62%)	2.44	(68%)	2.90	(65%)	2.88	(81%)

Date	Uncorrected D_LCO (mL/min/mmHg)		Corrected D_LCO (mL/min/mmHg)		Corrected KCO (mL/min/mmHg)	
09-12-2021 R	22.1	(80%)	21.9	(80%)	4.3	(92%)
24-08-2022 R	19.6	(72%)	19.2	(71%)	3.9	(82%)
14-02-2023 R	17.1	(63%)	17.0	(63%)	4.9	(103%)

Date	VC_{Pleth} (L)		RV_{Pleth} (L)		TLC_{Pleth} (L)		RV/TLC_{Pleth}	FRC_{Pleth} (L)	
09-12-2021 R	3.95	(81%)	1.43	(98%)	5.37	(85%)	27	2.69	(94%)
24-08-2022 R	3.86	(80%)	1.52	(105%)	5.35	(86%)	29	2.86	(101%)
14-02-2023 R	2.98	(62%)	1.45	(99%)	4.39	(70%)	33	2.33	(81%)

May

HRCT

Progressive increase in reticular markings in bases since 2021 study, involvement of apical segment of both lower lobes and very early changes in the posterior segment of left greater than right upper lobes. Changes would be consistent with reasonably rapid progressive silicosis.

July

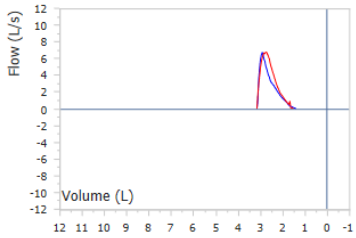
Symptoms: Patient feeling significantly worse in last 3 months, very short of breath even showering and dressing. Has developed widespread joint and muscle pain, swelling and tightness in hands significant progression since February.

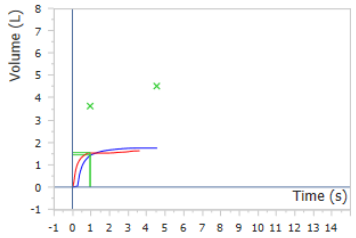
Lung Function

Further decline in all lung function parameters, nearly a 30% reduction in DLCO in 5 months

Forced Expiratory Volumes (BTPS) [10:02] GLI2012

		Pre BD	Post BD	%Change	Predicted Mean (Range)	Z-Score
FEV_1	(L)	1.52 (42%)*	1.63 (45%)*	7%	3.62 (>2.86)	-4.09
FVC	(L)	1.74 (39%)*	1.85 (41%)*	6%	4.49 (>3.55)	-4.69
FEV_1 /FVC		88%	88%		81% (>70%)	1.35
FEF_{25-75}	(L/s)	1.79	1.99	11%	3.54 (>2.01)	-1.67
FEF_{50} /FIF_{50}						





SUBDIVISIONS OF LUNG VOLUMES (Plethysmography) [10:26] GLI2021

		Observed	Predicted Mean (Range)	Z-Score
VC	(L)	1.94 (40%)*	4.83 (3.87-5.82)	-5.14
IC	(L)	1.04 (31%)*	3.41 (2.43-4.35)	-3.80
ERV	(L)	0.90 (71%)*	1.26 (0.55-2.22)	-0.76
RV	(L)	1.20 (81%)*	1.49 (0.84-2.27)	-0.70
TLC	(L)	3.14 (50%)*	6.31 (5.10-7.53)	-4.39
RV/TLC		38%	23% (15%-33%)	2.56
FRC	(L)	2.10 (73%)*	2.88 (1.97-4.04)	-1.39

SINGLE BREATH DIFFUSING CAPACITY [10:07] GLI2017

		Observed	Predicted Mean (Range)	Z-Score
$D_L CO_{uncorrected}$	(mL/min/mmHg)	10.1 (37%)*	27.13 (20.97-34.26)	-5.50
$D_L CO_{corrected\ for\ Hb}$	(mL/min/mmHg)	9.6 (35%)*	27.13 (20.97-34.26)	-5.72
$KCO_{corrected\ for\ Hb}$	(min/mmHg)	3.6 (77%)*	4.71 (3.68-5.82)	-1.72
V_A	(L)	2.6 (46%)*	5.80 (4.73-6.95)	-5.33
IVC	(L)	1.6 (35%)*	4.49 (3.55-5.45)	-5.21

HAEMOGLOBIN

		Observed	(Range)
Hb	(g/L)	165	(135-175)

SEQUENTIAL DATA

Date	Pre BD FEV_1 (L)	FVC (L)	FEF_{25-75} (L/s)	Post BD FEV_1 (L)	FVC (L)	FEF_{25-75} (L/s)
09-12-2021 R	2.86 (78%)	3.47 (76%)	2.85 (79%)	3.05 (83%)	3.75 (83%)	2.97 (82%)
24-08-2022 R	3.00 (83%)	3.76 (84%)	2.76 (78%)	3.08 (85%)	3.76 (84%)	3.29 (92%)
14-02-2023 R	2.36 (65%)	2.95 (66%)	2.20 (62%)	2.44 (68%)	2.90 (65%)	2.88 (81%)
11-07-2023 R	1.52 (42%)	1.74 (39%)	1.79 (51%)	1.63 (45%)	1.85 (41%)	1.99 (56%)

Date	Uncorrected D_LCO (mL/min/mmHg)	Corrected D_LCO (mL/min/mmHg)	Corrected KCO (mL/min/mmHg)
09-12-2021 R	22.1 (80%)	21.9 (80%)	4.3 (92%)
24-08-2022 R	19.6 (72%)	19.2 (71%)	3.9 (82%)
14-02-2023 R	17.1 (63%)	17.0 (63%)	4.9 (103%)
11-07-2023 R	10.1 (37%)	9.6 (35%)	3.6 (77%)

Date	VC_{Pleth} (L)	RV_{Pleth} (L)	TLC_{Pleth} (L)	RV/TLC_{Pleth}	FRC_{Pleth} (L)
09-12-2021 R	3.95 (81%)	1.43 (98%)	5.37 (85%)	27	2.69 (94%)
24-08-2022 R	3.86 (80%)	1.52 (105%)	5.35 (86%)	29	2.86 (101%)
14-02-2023 R	2.98 (62%)	1.45 (99%)	4.39 (70%)	33	2.33 (81%)
11-07-2023 R	1.94 (40%)	1.20 (81%)	3.14 (50%)	38	2.10 (73%)

Follow-up

Urgently referred to transplant team, Nintedanib prescribed and autoimmune testing requested.

<u>August</u>

Blood tests

Positive Antinuclear Antibody test (ANA) of 1.640, normal Erythrocyte Sedimentation Rate (ESR), positive Antineutrophil Cytoplasmic Antibodies (ANCA) which may be related to ANA, negative Extractable Nuclear Antigen (ENA) antibodies panel, Rheumatoid Factor was 26, Angiotensin Converting Enzyme (ACE) normal and C-Reactive Protein (CRP) normal. Patient was referred to Rheumatology for investigation.

<u>September</u>

HRCT

Possible Usual Interstitial Pneumonia (UIP) pattern of interstitial lung disease with no honeycombing.

<u>October</u>

Rheumatology who diagnosed scleroderma via skin biopsy, patient started Mycophenolate. Prescribed supplementary oxygen due to desaturation with exertion.

December

Lung Function

Parameters stabilised.

SEQUENTIAL DATA

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09-12-2021 R	2.86 (78%)	3.47 (76%)	2.85 (79%)	3.05 (83%)	3.75 (83%)	2.97 (82%)
24-08-2022 R	3.00 (83%)	3.76 (84%)	2.76 (78%)	3.08 (85%)	3.76 (84%)	3.29 (92%)
14-02-2023 R	2.36 (65%)	2.95 (66%)	2.20 (62%)	2.44 (68%)	2.90 (65%)	2.88 (81%)
11-07-2023 R	1.52 (42%)	1.74 (39%)	1.79 (51%)	1.63 (45%)	1.85 (41%)	1.99 (56%)
18-09-2023 R	1.63 (45%)	1.87 (42%)	2.24 (63%)			
16-10-2023 R	1.54 (43%)	1.67 (37%)	2.24 (64%)	1.55 (43%)	1.68 (37%)	2.69 (76%)
19-12-2023 R	1.70 (47%)	1.86 (41%)	2.55 (72%)			

Date	Uncorrected D_LCO (mL/min/mmHg)	Corrected D_LCO (mL/min/mmHg)	Corrected KCO (mL/min/mmHg)
09-12-2021 R	22.1 (80%)	21.9 (80%)	4.3 (92%)
24-08-2022 R	19.6 (72%)	19.2 (71%)	3.9 (82%)
14-02-2023 R	17.1 (63%)	17.0 (63%)	4.9 (103%)
11-07-2023 R	10.1 (37%)	9.6 (35%)	3.6 (77%)
18-09-2023 R	8.7 (32%)	8.3 (31%)	3.2 (68%)
16-10-2023 R	8.8 (33%)	8.3 (31%)	3.9 (82%)
19-12-2023 R	8.1 (30%)	8.2 (30%)	3.1 (67%)

Date	VC_{Pleth} (L)	RV_{Pleth} (L)	TLC_{Pleth} (L)	RV/TLC_{Pleth}	FRC_{Pleth} (L)
09-12-2021 R	3.95 (81%)	1.43 (98%)	5.37 (85%)	27	2.69 (94%)
24-08-2022 R	3.86 (80%)	1.52 (105%)	5.35 (86%)	29	2.86 (101%)
14-02-2023 R	2.98 (62%)	1.45 (99%)	4.39 (70%)	33	2.33 (81%)
11-07-2023 R	1.94 (40%)	1.20 (81%)	3.14 (50%)	38	2.10 (73%)
16-10-2023 R	1.75 (36%)	1.26 (85%)	3.01 (48%)	42	1.89 (66%)

Future Follow-up

Rapid progression of lung disease may be in response to development of scleroderma. Referred for IV Cyclophosphamide, monthly, for 6 months with hopes of slowing progression, and still actively being reviewed for transplant assessment.

Conclusion

The patient characteristics in this case study support the findings from the recent review by Hoy et al[2]; this patient he was 41 years of age, an immigrant and worked with ES for 12 years.

It also demonstrates the severe impact that ES-induced silicosis can have on a person's life due its short latency and rapid deterioration of disease. Hopefully, other countries will follow Australia's lead in banning the use of the product or at least impose strict regulations to workplaces to protect workers in the future.

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