



# SILICOSIS AND ITS ASSOCIATION WITH AUTOIMMUNITY: THREE CASES OF SILICOSIS-RELATED ANCA VASCULITIS



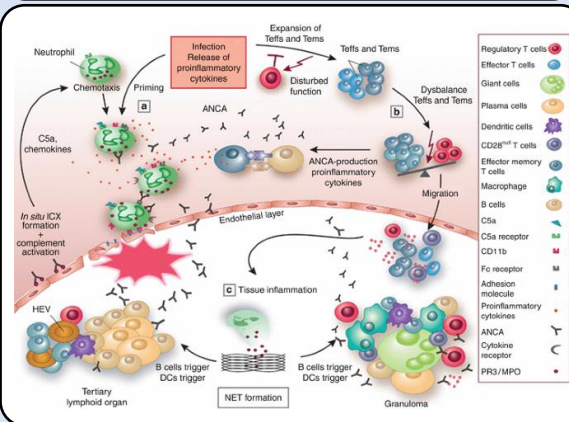
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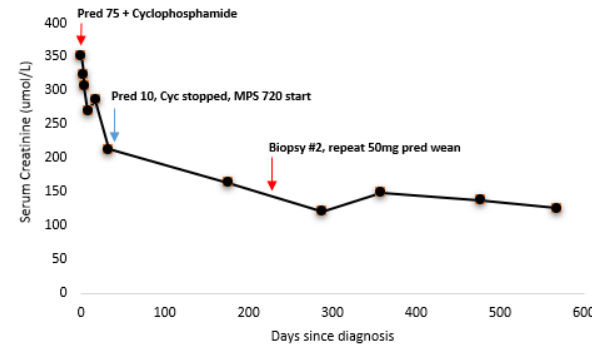
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## Background

- Growing interest in the association between Silicosis and development of ANCA-vasculitis
- Silicosis = fibrotic lung disease, developing from occupational inhalation of respirable crystalline silicon dioxide particles in construction, mining and automobile repairs (1).
- *In vitro* studies suggest dysfunctional immune responses to silica contribute to development of a variety of autoantibodies including ANCA (2).
- We present three cases of middle-aged men with Silicosis who developed ANCA vasculitis following their diagnosis.



## Case 1 – MPO vasculitis with IgAN



### 58-year-old male stone mason with silicosis for 8 years.

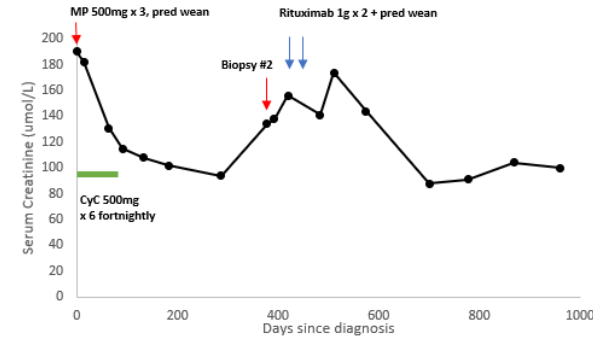
- Sx: Haemoproteinuria with 1/12 malaise, nocturia.
- PMHx: Silicosis, Resected melanoma, nil smoker
- Ix Cr 352, eGFR 16, uMCS <10/>500/20, uACR 210, MPO-ANCA 550, pANCA 640, others –ve.

#### Biopsy: ANCA-vasculitis with IgA nephropathy.

- 26/9/22: 26 glom, 14 fibrocellular, 5 cellular. IF 4+ granular IgA. 3+ IgM/C3/Iamda. EM: neg.
- 16/5/23: 15 glom, 3 fibro cellular, 2 cellular.

**Treatment:** 75mg prednisolone wean, CYC 3 month with oral treatment. Changed to MPS 720mg BD.

## Case 2 – MPO vasculitis with IgAN



### 41-year-old male retired stone mason with silicosis for 10 years.

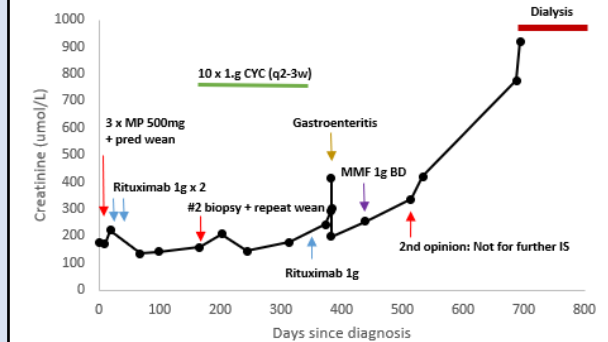
- Sx: Haemoproteinuria with Cr 190. Vasculitic rash.
- Retired stone Mason with **silicosis**. Non-smoker
- PMHx: Silicosis. Nil meds. NKDA, non-smoker
- Ix Cr 184, eGFR 39, uMCS 80/>500/<10, uACR 200, MPO-ANCA 2.1, ENA +ve, anti-Ro52 +ve

#### Biopsy: ANCA-vasculitis with IgA nephropathy.

- 8/12/2021: 25 glom, 11 cellular, 5 fibrocellular. IF 2 + IgA and IgM mesangial. EM = DFPE.
- 19/01/2023: 14 glom, 5 fibrocellular+1 cellular.

**Treatment:** 30mg prednisolone, CYC 6xIV 500mg. Change to MPS 720mg BD, then rituximab 2 x 1g induction + rpt. wean

## Case 3 – MPO vasculitis + IC-GN



### 38-year-old male stone mason with silicolymphadenopathy.

- Sx: Mild abdominal pain, haemoproteinuria moved from Darwin with Cr ~130 umol/L. Ex-smoker.
- PMHx: HTN, obesity, prev. cocaine/amphetamine.
- Ix: Cr 176, eGFR 41 uMCS 20/500/<10, uACR 220 MPO-ANCA >740, pANCA >2560, ESR 70.

#### Biopsy: ANCA-vasculitis with immune-complex GN.

- 11/11/2020: 6 glomeruli, 2 fibrocellular/fibrous crescents. 20% IFTA. IF weak IgG.
- Biopsy #2 similar.

**Treatment:** 60mg prednisolone and 2 x rituximab 1g, cyclophosphamide 10 x 1.2g IV. Change to MMF 1g BD.

## References

1. Leung et al. 2012, *The Lancet*, 379(9830):2008-18.
2. Nishimura et al. 2017, *J Gen Fam Med*, 18(5):288-90.
3. Gomez-Puerta et al. 2013, *Autoimmune review*, 12(12): 1129-35.
4. Tervaert 2013, *Encyclopedia of Metalloproteins: Springer (image)*

## Take away points

- Patients with silica ANY silica exposure have a 3 x higher likelihood of developing ANCA vasculitis (2, 3).
- Further work needed to explore natural history and treatment characteristics of silica-related ANCA-vasculitis and mechanisms of auto-antibody development.
- Patients with prev. silica exposure should be considered for urinary testing and autoimmune profiling for earlier diagnosis and treatment.