Protracted Katayama Syndrome

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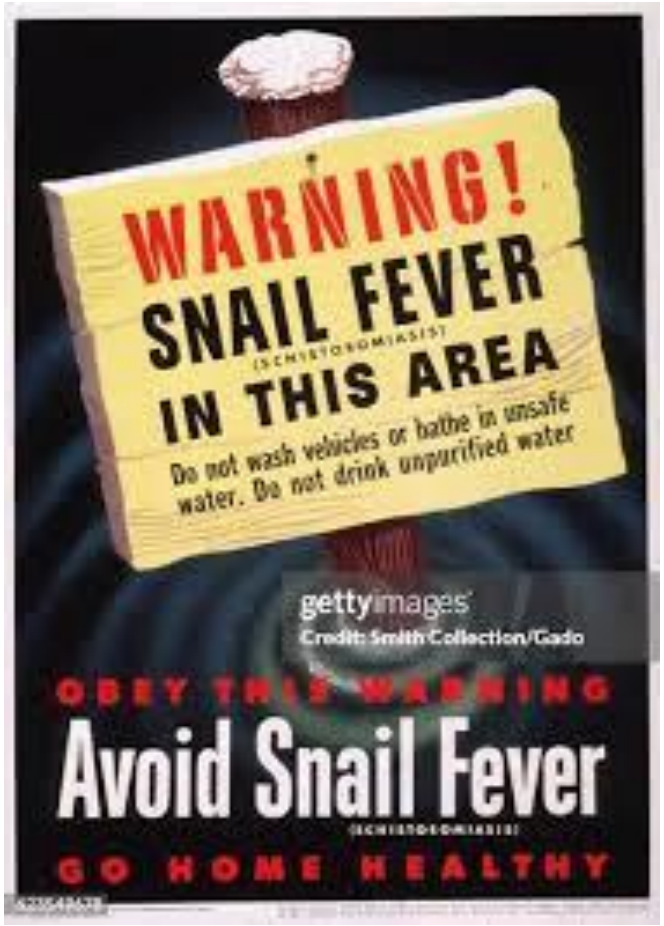
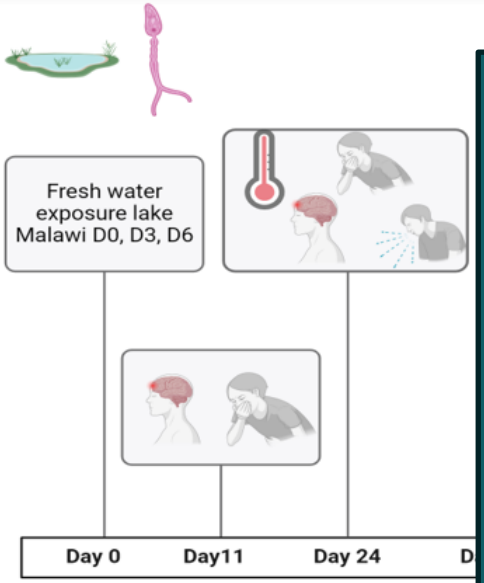


Boy 17 year with fever, cough and general body pains after a trip to Malawi

- 15q13.3 microdeletion syndrome autism spectrum disorder
- A genetic variant in TACI (TNFRSF13B), predisposing him to common variable immunodeficiency (CVID)
  - IV immunoglobulins from the age of 2 to 12 years due to frequent upper respiratory and urinary tract infections
  - Restarted preventively during the COVID-19 pandemic until 15 months before the current episode

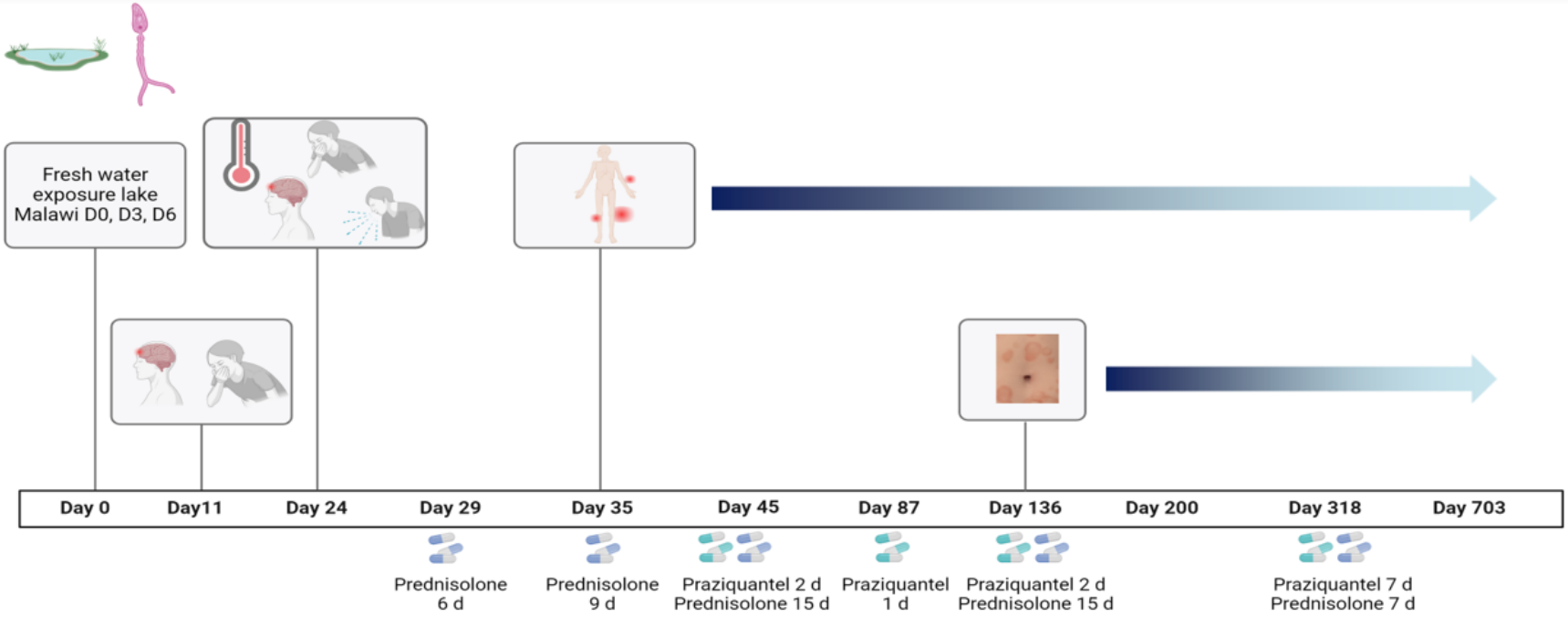
History

- 3 Weeks Malawi with 2 friends
- First two weeks: hiking and safari
- Last week: the group swam three times in Lake Malawi (Cape Maclear)
- Other exposures: drinking from a stream and eating raw vegetables and fruit, brushing teeth with tap water
- Two of the three friends: fever, headache, myalgia, cough, urticaria and angioedema


Fresh water exposure lake Malawi D0, D3, D6
Day 0
Day11
Day 24

Further findings

- 38°C
- Deep palpation of the abdomen slightly tender, liver edge palpable
- There were crusts on the lips but no skin rash
- Lab:
  - Eosinophilia 660 cells/µL
  - Platelets were 113,000 x 10^9/L, CRP 52 mg/L
  - Leptospirosis RDT weakly positive
    - ceftriaxone and oral doxycycline
  - MAT on paired sera: neg
  - Rise in eosinophil count (up to 2,030 cells/µL)
    - prednisolone 32 mg for 6 days D4 hospitalization for a tentative diagnosis of Katayama syndrome
- Quick improvement
- But day after stopping prednisolone, pain in left forearm and knees
  - Eosinophil count 4,250 cells/µL
    - Prednisolone restarted 9 days in a tapering dose.



	D 0	D 11	D 24	D 29	D 35	D 45	D 87	D 136	D 200	D 318	D 703
Eosinophils (eg's/µl)			660	2030	4250	1030	590	560	340	790	710
PCR Dra1 (CT)			0	46,01		40,22	36,12		33,59	41,34	Neg
PCR Sm 1-7 (CT)			0		0	0					
ELISA (nl. < 1.00)						0,86 (neg)	1.07 (pos)	0,63 (neg)	0,51 (neg)	0,24 (neg)	0,16 (neg)
IHA						Neg	Neg	Neg	Neg	Neg	Neg
CAA						Pos		Neg		Neg	

Lessons learned

- In hindsight: the persisting symptoms and laboratory abnormalities were likely unrelated to the *Schistosoma*
- Troublesome interpretations: immunocompromised status, persistence of features suggestive of Katayama syndrome and fluctuating/decreasing Dra1 Ct values PCR
  - -- > additional testing and repeated treatments
- *Schistosoma* DNA in blood demonstrated: high Se and Sp for early D/ acute schisto
- Remains positive for many months after treatment < -- > PCR on stools or urine
- PCR on serum: not necessarily reflect the persistence of living worms
  - test of cure?

- What can be translated from other studies to immunocompromised?

Lessons learned

- CAA most reliable
- UCP-LF CAA in batch twice a month in LUMC only
- Shipping international samples: logistics and cost
- Delays ++

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IHA						Neg	Neg	Neg	Neg	Neg	Neg
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Lessons learned

- Serology: Se both serological assays suboptimal for *S. haematobium* infection
- Impaired antibody production in our patient??

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Immunologist consultation

- Immune response to schisto mostly Th 2 mediated + IgE, eosinophils and macrophages
- < -- > Suceptibility to reinfection: ~ parasite-specific blocking IgG4 immunoglobulins, counteracting IgE
- Humoral immunodeficiency unclear in our patient
- Was he immunodeficient? Or was this transient?

- Schisto common: but no reports link inborn errors of immunity with a potential predisposition to schistosomiasis or persistent Katayama syndrome
  - Underreporting in endemic regions?


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