PIMS-TS and Sickle cell disease Paediatric Rheumatology perspective

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Rheumatology perspective

- Challenges
- Paediatric Rheumatology role in PIMS –TS at Evelina
- Evolving clinical case
- Reflection
- Future

First impressions…

Patient brought to our attention in a MDT meeting

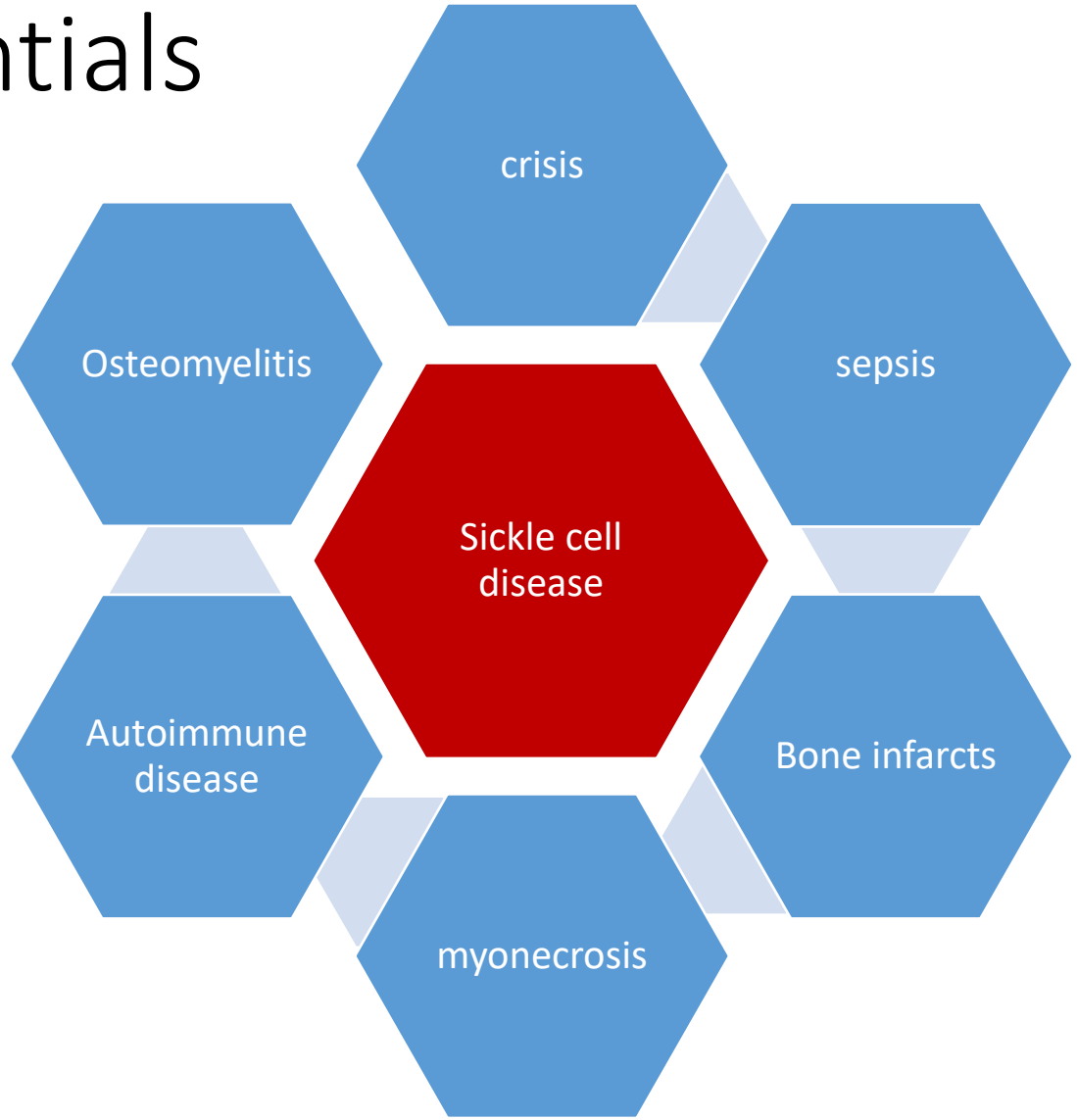
First reaction- speak to Baba (Inusa) myself

On the same day discussed twice to be absolutely sure

Clinical picture and lab markers suggestive of PIMS-TS

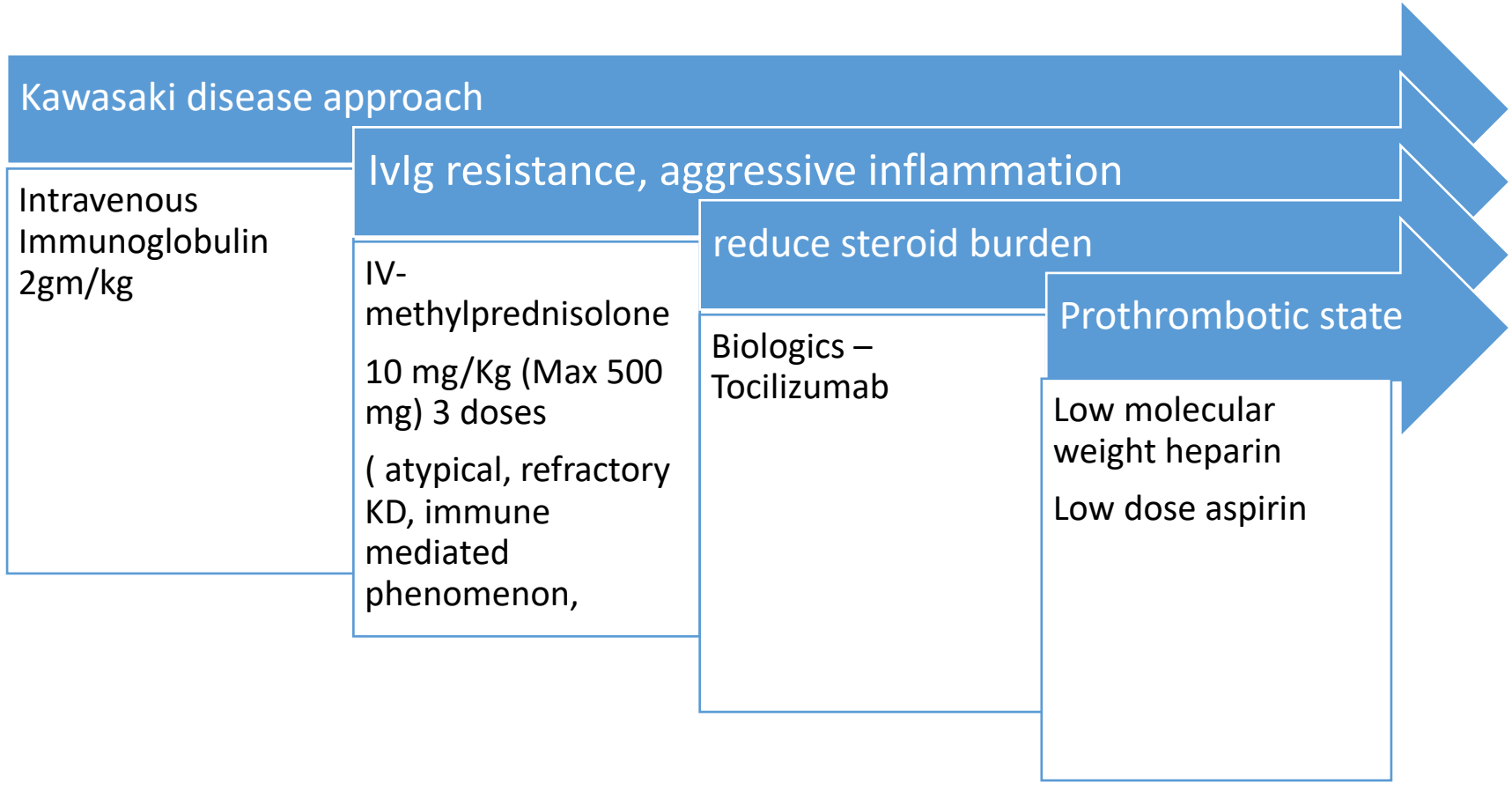
But, it is a diagnosis of exclusion

Differentials



Hughes et al.Haemoglobinopathies and the Rheumatologist.Rheumatology (Oxford) . 2016 Dec;55(12):2109-2118.

Therapeutic approach



Progress

- CRP >200 and high ferritin not explained by other causes
- Fever noticed 2-3 days after the initial presentation.
- High fibrinogen and D- dimers unaccounted
- But, good response to the combination treatment
- Fever settled and CRP trending down, but coronary complications noted
- After 3 days of IV methylprednisolone switched to Oral prednisolone

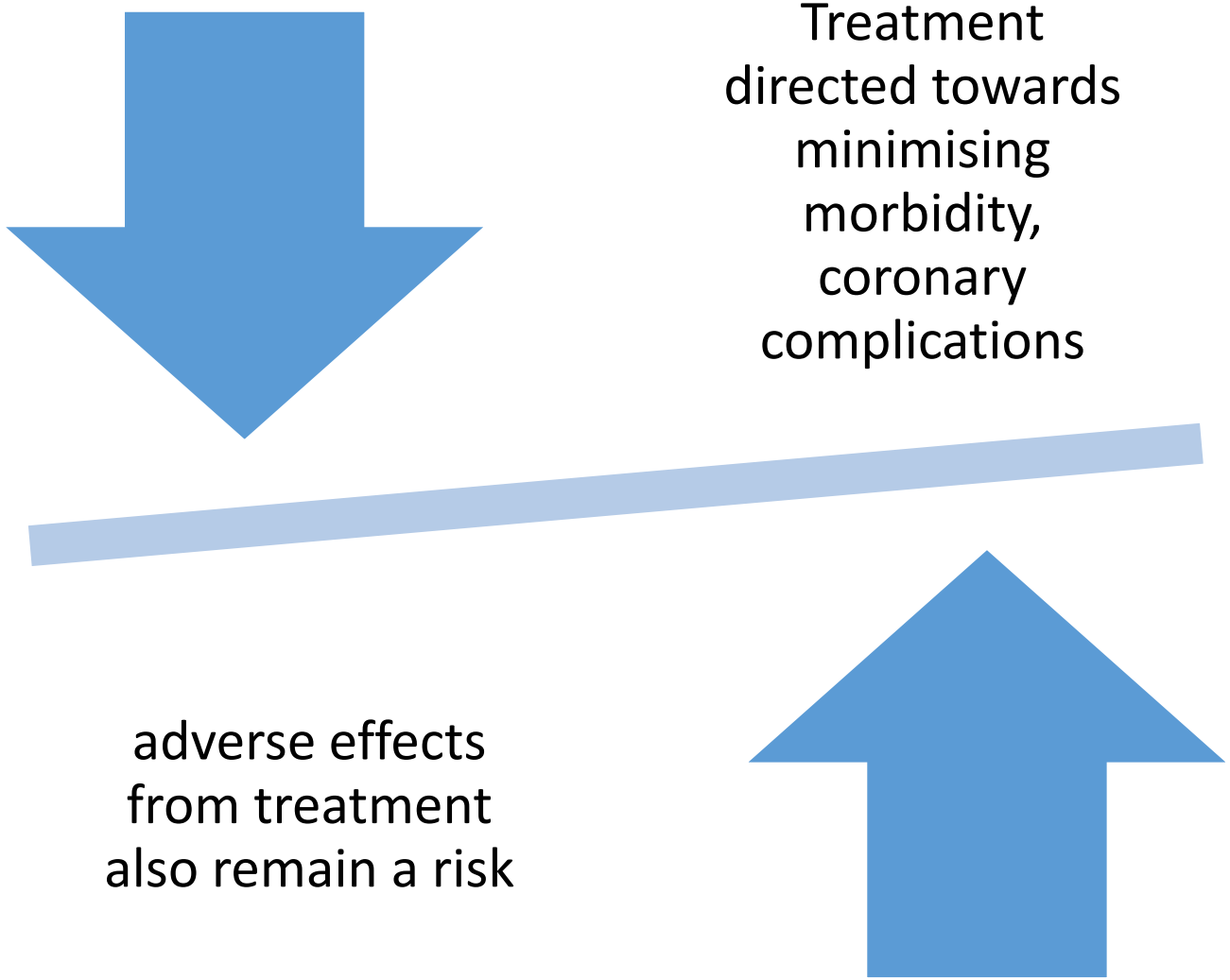
(60 mg)

- Still concerns about “thigh pain”- MRI requested
- Chest pain – CT pulmonary angiogram normal
- Cardiac findings don’t explain the chest symptoms
- Diagnosis of PIMS-TS challenged all along, until cardiac findings on CT confirmed
- But things changed…

Reflection

PIMS –TS is a hyperinflammatory, prothrombotic condition

How do we get the balance right?



- IvIg- Thromboembolism, hyper viscosity syndrome
- Steroids – precipitate vaso-occlusive crises (VOC) , or rebound VOC
- Steroid induced hypertension, Posterior reversible encephalopathy syndrome

Guidelines for medical treatment of acute Kawasaki disease: Report of the Research Committee of the Japanese Society of Pediatric Cardiology and Cardiac Surgery (2012 revised version). Pediatrics International (2014) 56, 135–158

Darbari et al. Severe Vaso-Occlusive Episodes Associated with Use of Systemic Corticosteroids in Patients with Sickle Cell Disease. J Natl Med Assoc. 2008 Aug; 100(8): 948–951.

Couillard S et al. Steroid treatment in children with sickle-cell disease.Haematologica. 2007 Mar; 92(3):425-6.

- Intrinsic vessel abnormality of the vessels – particularly CNS
- Hyper inflammatory state and hypercoagulatory state in PIMS-TS, and sickle cell prothrombotic state- a double whammy?
- Did aspirin and LMWH have a role?

Future implications

- What can we learn from this
- IvIg and steroids? Beneficial or harmful?
- SCD and autoimmune conditions (SLE and polyarthritis) – biologics well tolerated.
- Biologics early on with lowest possible dose steroids
- Aspirin and LMWH?

- Thank you