PYREXIA OF UNKNOWN ORIGIN

A DIAGNOSTIC DILEMMA

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HISTORY

- 4 years old/female child
- Developmentally normal
- Immunised for age
- No significant past illness

- H/O intermittent fever –1 month
- No other localising symptoms
- Had received multiple oral antibiotics

EXAMINATION

- Febrile, coated tongue.
- Abdomen – mild hepatomegaly.

- TC – 3100 (N 17,L 78), Hb – 9.4, Plat- 2.4L
- ESR - 40 mm/hr
- SGOT – 69 : SGPT – 19.
- Blood culture - sterile.

TREATMENT

- Partially treated enteric fever - 5 days of IV Ceftriaxone
- She was afebrile for 24 hours
- Discharged on oral antibiotic (Cefixime) for a total duration of 2 weeks
- Plan was to repeat blood counts at review

FOLLOW UP

- Completed course of oral cefixime
- Persisting fever spikes – 8 weeks duration

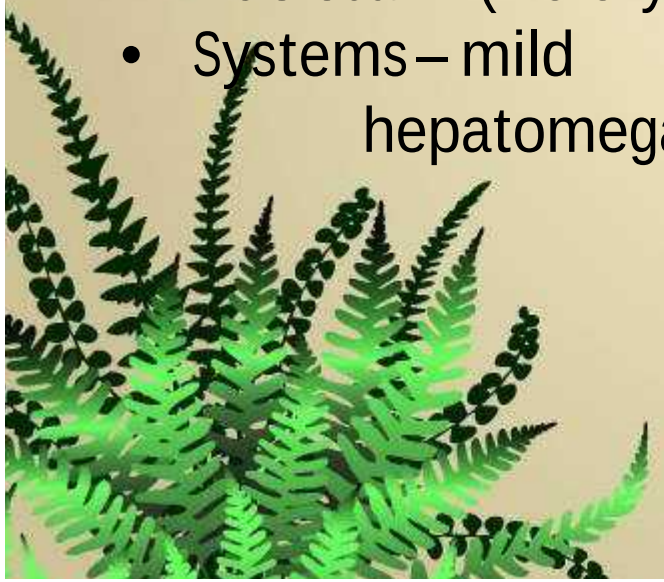
- Repeat CBC - Persisting Leukopenia
  (TC – 3100, Hb - 9.4, Plat – 2.4 L)
- Hence she was readmitted for further workup

HISTORY - RELOOK

- H/O poor appetite – even before the illness
- No skin rashes/joint pain/joint swelling
- No H/O loss of weight
- No pallor / clinical bleeds
- No H/O contact with TB
- No H/O recent travel/contact with pets
- No H/O any drug intake
- No H/O surgery in past
- Vegetarian by diet – no H/O intake of any raw food

EXAMINATION

- Febrile , not sick looking
- Pallor +
- Painless Bilateral upper eyelid swelling was seen
- Glossitis +. No oral ulcers
- No significant lymphadenopathy
- No skin rash/joint swelling/bone tenderness
- BCG scar + (No erythema)
- Systems – mild hepatomegaly



DIFFERENTIAL DIAGNOSIS

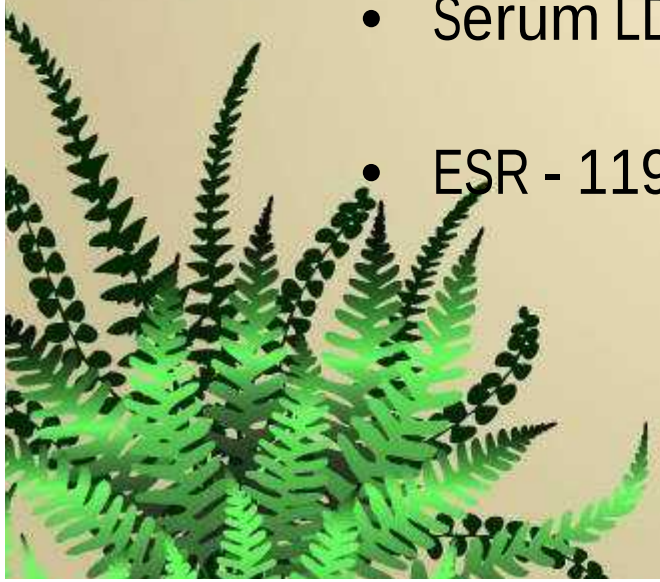
- Prolonged viral illness (EBV)
- Enteric fever
- Tuberculosis
- Collagen vascular disease
- Evolving malignancy
- Infection associated HLH
- Immunodeficiency – HIV

WORK UP

- TC - 3,500 cells
- DC - N 40/75, L 34/75, M 01/75
- Hb – 7.6 g/dl
- Platelet – 3.3 lakhs

- Peripheral smear – normocytic normochromic anemia, no evidence of hemolysis, no abnormal cells
- DCT – negative, Reticulocyte count - 1 %
- Serum LDH – 3740 (elevated)

- ESR - 119 mm/hr



- Renal function test - normal

- SGOT - 105 IU/L
- SGPT - 26 IU/L
- Serum albumin – 3.4 g/dl

- ECHO (to rule out Kawasaki disease / infective endocarditis) – Normal

Work up for infective etiology

- EBV VCA IgM – negative
- WIDAL – negative
- Blood & Bone marrow culture – sterile
- Urine microscopy & culture – normal
- CSF analysis was normal

- Scrub typhus IgM – Non reactive
- HIV ELISA - Non reactive

Work up for TB

- ESR – elevated (119mm/hr)
- Mantoux – negative
- CXR – normal
- CT chest – normal. No mediastinal nodes

Work up for malignancy

- Peripheral smear – no abnormal cells
- Bone marrow smear – reactive marrow.
  No atypical cells.
- USG abdomen – mild Hepatosplenomegaly
- Serum LDH – 3740

Autoimmune workup

- ANA – negative
- ds DNA - negative

Work up for infection associated HLH

- Ferritin – 898
- TGL – 329
- Fibrinogen – 375

Management

- Temperature monitoring
- NSAID - Ibuprofen
- Empirical doxycycline (to cover any atypical organism)

Ophthalmologist consult

‘BILATERAL DACRYOADENITIS’

?ORBITAL PSEUDOTUMOUR

underlying autoimmune disease or chronic infection

Advised CT Orbit/oral steroid

DACRYOADENITIS

- Inflammation of the lacrimal gland
- Acute or chronic
- Acute – Viral / Bacterial
- Chronic – Inflammatory disorders
- C/F: Painful swelling in the region of upper eyelid, fever, watering of eyes
- Inv: CT/MRI , Biopsy
- Treatment: Rest, warm compresses

ORBITAL PSEUDOTUMOUR

- Idiopathic, nonmalignant orbital inflammation
- Etiology – Not known

  Autoimmune etiology
- Present as a painful, unilateral orbital swelling
- Inv: MRI, biopsy
- Treatment: High dose steroids, immunosuppression, radiotherapy.

THE FINAL CALL

- Parents were given the option of further workup – CT orbit and proceed or to start the child on steroid and look for the response.

- "NO" – For further evaluation

- She was started on oral steroids.
- She was afebrile for 2 days in the hospital.
- Discharged on steroids.

FOLLOW UP

- She was afebrile.
- Eyelid swelling started resolving.

Investigations at follow up

2 weeks after discharge

- TC – 8300 cells/cu.mm, Hb – 7.6g/dl
- Platelet count - 6.4Lakhs/cu.mm
- ESR – 108 mm/Hr
- ECHO (? Kawasaki disease) - normal

Clinical diagnosis

? INCOMPLETE KAWASAKI DISEASE

- Started her on antiplatelet dose of aspirin.
- Advised to review in 2 weeks time.

At review

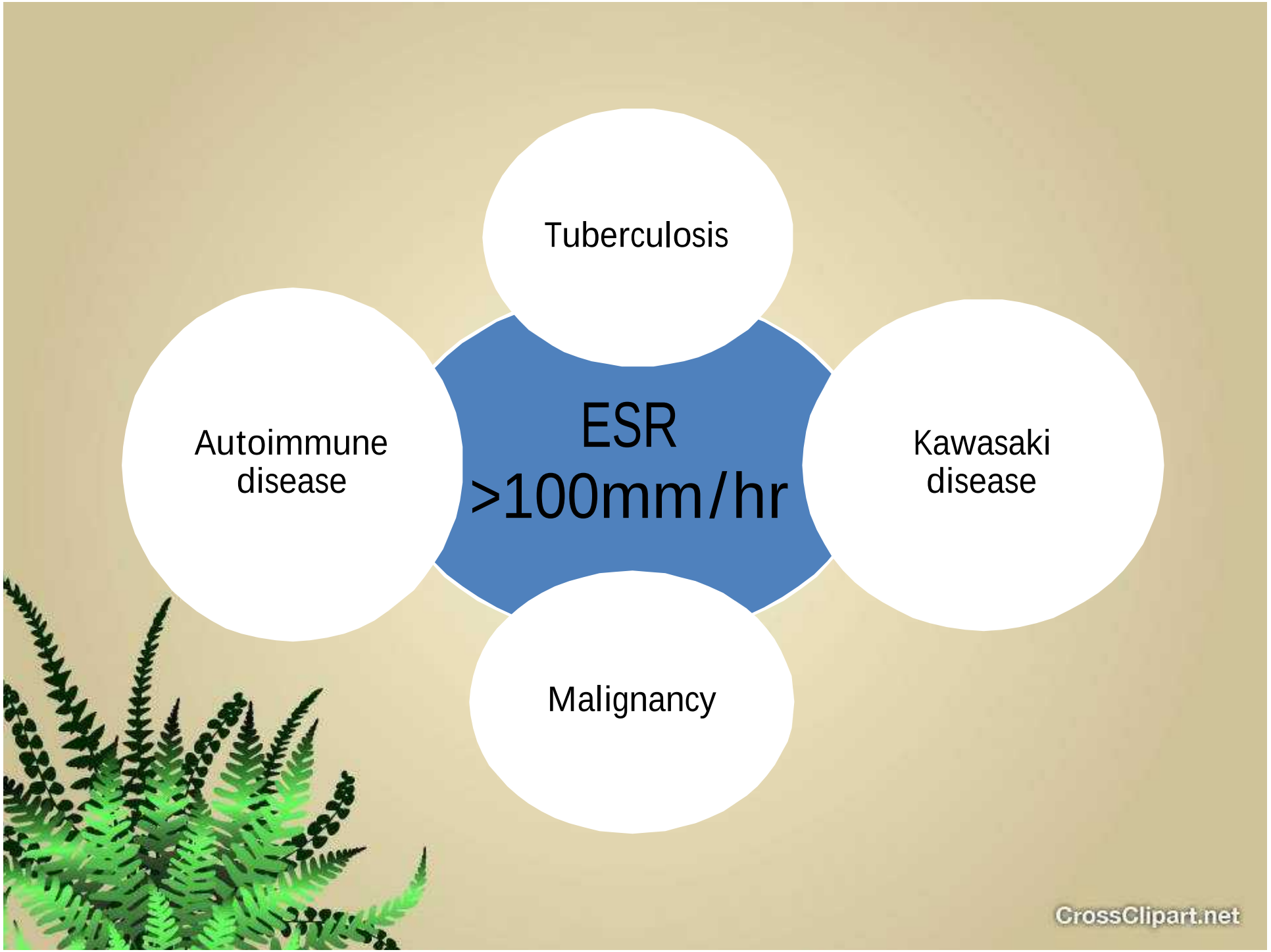
- Clinically afebrile.
- No new signs or symptoms.

<u>Repeat investigations:</u>

- TC 6600, Hb 10, Platelets 3.3 L
- ESR -24 mm/Hr
- CRP was negative

Possibilities:

- Can still be an incomplete KD, autoimmune disorder or evolving malignancy.
- She needs frequent follow up and re-examinations to make a definitive diagnosis.


Tuberculosis
Autoimmune disease
ESR >100mm/hr
Kawasaki disease
Malignancy
CrossClipart.net

Baseline

- CBC, ESR, CRP, Peripheral smear
- Mantoux
- Bone marrow smear
- Cultures – blood, urine, CSF, bone marrow

Serology

- Infectious mononucleosis, brucellosis, enteric fever, rickettsial, leptospirosis, lyme's disease
- ANA, dsDNA, ANCA

Imaging

- Chest x-ray, USG Abdomen, CT chest, whole body MRI, PET
- Bronchoscopy, laparoscopy, mediastinoscopy, GI scopy

TAKE HOME MESSAGES

- Be rationale in investigating a child with PUO.
- Don’t be in a hurry to label the child with a diagnosis.
- Counsel the family.
- YOUR PATIENCE IS ESSENTIAL.