A child with sickle cell and PIMS-TS

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10 year old boy in the Emergency Dept

Presentation

2/7 Pain back, chest, thigh

Afebrile on arrival

No cough/D+V/dysurea/ muco-cutaneous change

Off food/drink

No unwell contacts

Background

Sickle cell anaemia (ss)

Takes hydroxyurea, PenV, folic acid, abidec

Previous painful crises
Enuresis ongoing
Epistaxis 2019
Osteomyelitis 2014

Initial assessment

8/10 pain, obs normal

Examination normal

Hb 100g/L (baseline) Plt 145 x10^9, Lymph 2.1, Neutr 6.6, CRP 6mg/L,

WCC, U+Es, LFTs normal

CXR normal

Sickle crisis

+ Difficult pain management

Pain management

Encourage oral hydration

Consider imaging if change

Monitor for fever

Progression of illness

D1-2

Pain management

Requiring PCA & clonidine

D 3

Fever ?Source

HR rising; prior CRP rising 63

Abdomen pain

Commenced Ceftriaxone

+ Increased macrogol

D4

PIMS-TS?

Fever 38.5

Abdomen pain persists

Thigh/chest/back pain

Examination:

Abdomen tender LIF

Tender hip but full ROM

Investigations

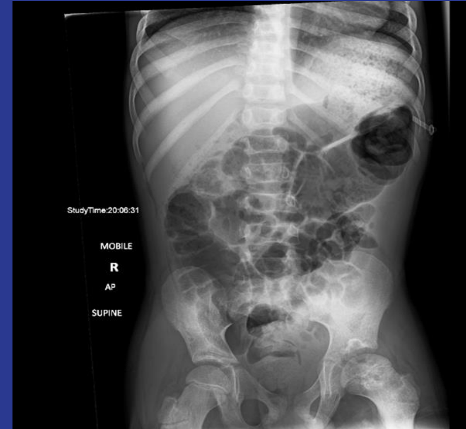
Virology
Covid PCR neg; serology positive [later]

Bacteriology
Blood + urine culture negative

Cardiology
ECG normal, ECHO emerging coronary changes

Bloods:
Hb 88 g/L
WCC 11.4 Lymph 0.4,Neutr 7.3x10^{9},
Ferritin 880 ug/L
Ddimer 8.5 mg/L
Na 133 mmol/L
Albumin 39 g/L
CRP 266 mg/L

Xray abdomen: constipation



Case definition (RCPCH)

1. Persistent fever, inflammation, organ dysfunction

2. Exclusion of other microbial cause

3. +/-SARS-CoV-2*

Additional features

Lethargy and Myalgia
Abdominal pain/Diarrhoea/Vomiting
Rash/Conjunctivitis
Hypotension
Raised CRP
Raised Ferritin (>500)
Lymphopenia / Neutrophilia
Raised Fibrinogen
Raised D-Dimer
Platelets initially low or normal
Raised Troponin and B-NP
Renal dysfunction

***Not present at time**

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***Not present at time**

1.PIMS TS

2.Sickle crisis
+Painful thighs

3.Sepsis?

4. Abdominal pain

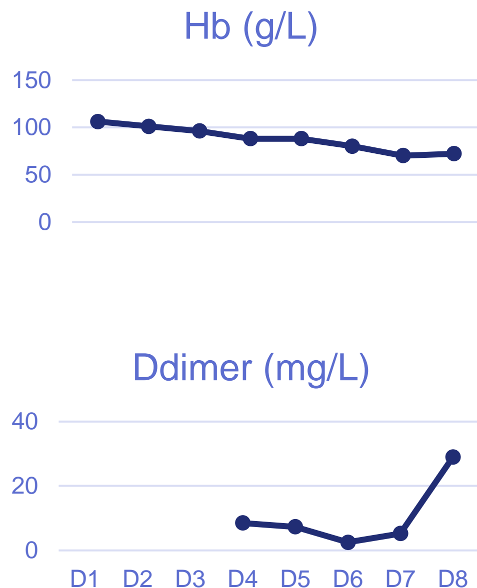
1. IVIG 2g/kg
Methylpred 10mg/kg
Tocilizumab D2 of diagnosis
Aspirin LD
Dalteparin
Colecalciferol 100,000 IU

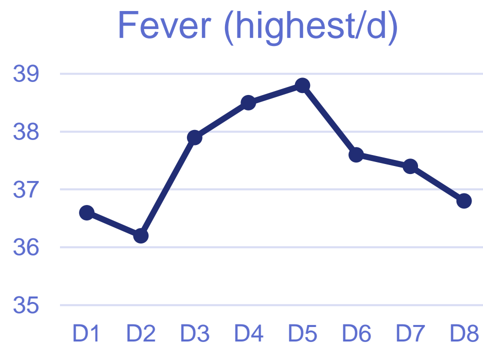
2. Analgesia
Hydroxyurea

3. Continue ceftriaxone

4. Omeprazole
Laxatives increase Movicol BD
Surgical review [nil concern]

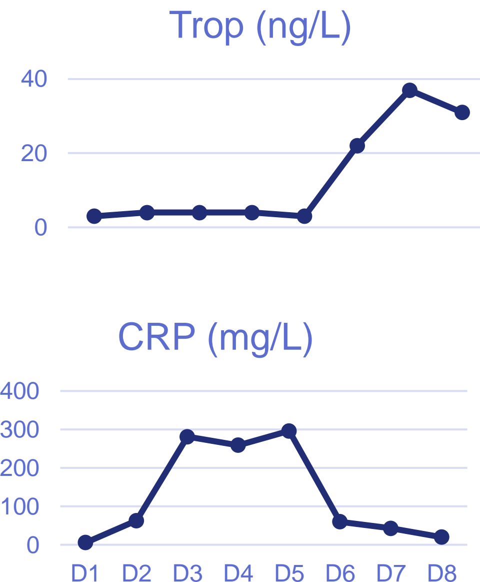
Investigations and progress





Methylpred
IVIG

Tocilizumab



Progression of illness

D5-D6

Apyrexic 24hrs

MRI showed chronic avascular necrosis + bone infarcts not septic

Epistaxis >20mins Hb 70

D7

Chest pain + palpitation

Twave inv ECG leads 1-3

Trop & BNP rise

CTPA - no PE

Coronary dilation

D8 morning

Abdo + leg pain persistent

Blood transfusion

PIMS TS

- Coronary artery dilatation
- Chest pain/ palpitations
- Sickle crisis
- Painful thigh
- Epistaxis with Hb drop
- Sepsis?
- Abdominal pain / constipated

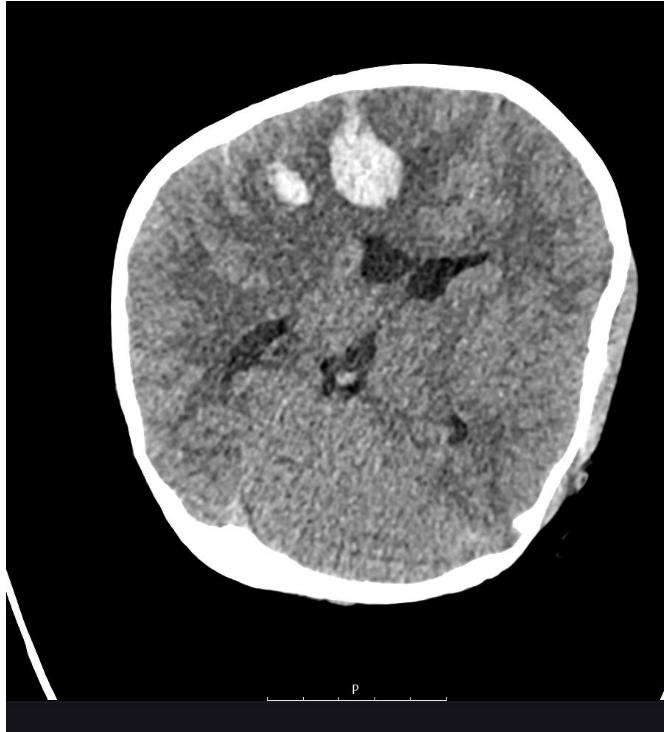
D8 Clinical Deterioration

- Drowsy >> lower GCS
- New onset headache + vomiting
- Hypertension
- New Left facial droop
- New Left sided weakness in upper and lower limbs

Impression and Initial Management

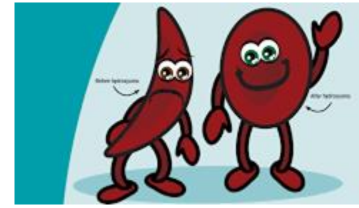
- ? Raised ICP secondary to stroke
- Neuroprotective measures including intubation
- Transferred arranged to PICU
- Start Levetiracetam
- Stop Dalteparin, aspirin and sedation

CT Brain



Rt sided Haemorrhagic stroke frontal lobe with midline shift

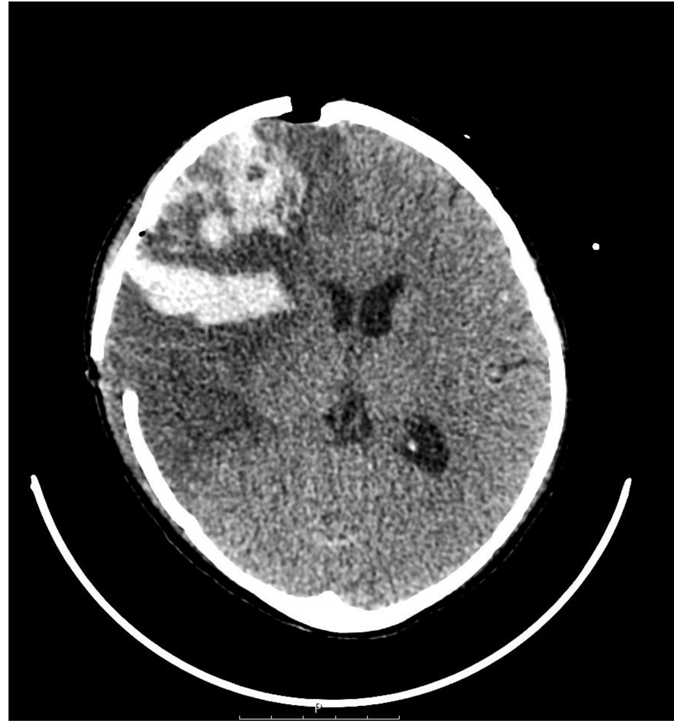
Current situation



Update from Kings' 13/06

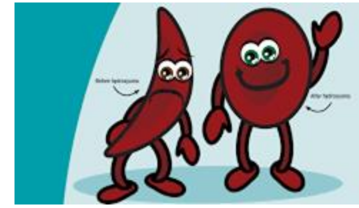
Craniotomy, surgical evacuation, ICP monitoring
Transfused peri-operatively
Slow improvement in GCS off sedation
ACA + MCA infarction on CT head
Extensive saphenous vein+ IVC thrombus >> IVC filter
Spiking temperature with raised CRP: IV Pip-taz, off steroids

CT Brain



Infarcts ACA/ MCA region

Conclusion



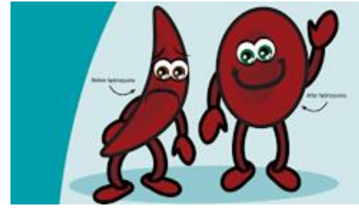
Difficult case

10 yo with sickle crisis

Addition to + complicated by

- PIMS-TS
- Haemorrhagic stroke
- Extensive thrombi

Challenges



Difficult questions

PIMS vs sepsis vs sickle crisis

Abdominal pain differentials

Difficulties in anticoagulation

Sickle management in Covid and PIMS-TS illness

Thanks!

+ Any questions?

Acknowledgements:

- Paediatrics
- Haematology
- PID
- Rheumatology
- Cardiology
- PICU
- Nursing team

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