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for rare or low prevalence
complex diseases

Network

Paediatric Cancer
(ERN PaedCan)



16 Sept 2026

Presenter: Alexandra Norman

Expert: Leo Kager

DYING FOR A DIAGNOSIS: SUCCESSFUL TREATMENT OF TAFRO SYNDROME IN A CHILD

Moderation: Andishe Attarbaschi





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COI declaration

- Nothing to declare



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Case report- acute presentation

9 year old girl

Previously fit and well

Short history
abdominal pain
diarrhoea
intermittent fever
nose-bleeds



Widespread oedema

Pleural effusions

Ascites

Splenomegaly

Renal failure

Low albumin



HDU support





Case report

CT chest/abdomen-
Enlarged lymph nodes in
abdomen, max 14mm in
diameter, splenomegaly

Normal immunoglobulins
and other immunological
markers negative

Ascitic fluid:
inflammatory
cells only

Bone marrow/trephine
biopsy: hyper cellular,
non- fibrotic marrow
with increased
myeloid-to-erythroid
ratio and
dysmegakaryopoiesis

INVESTIGATIONS

Whole body
MRI/US: no obvious
target for biopsy

PET: moderately
active lymph nodes,
more obvious in
abdomen

Ferritin
266

Creatinine 119
> 198 $\mu\text{mol/L}$

sCD25 > 320

Viral screens and
blood cultures
negative

CRP 150

Hb 76 g/L

Platelets 116 $10^9/\text{L}$





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Case report- new bruising/bleeding



Platelets: $96 \cdot 10^9/L$

PT 15.2 s. (10.6-14.9)

APTT: 71.2 s (24 - 35)



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What has caused the significant bruising?

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- Thrombocytopenia
- Acquired haemophilia
- Disseminated intravascular coagulation (DIC)
- Acute renal dysfunction





Acquired haemophilia A



- anti-FVIII antibodies
- Two previous children with acquired haemophilia and probable TAFRO have been reported
- Treatment with porcine FVIII and subsequently recombinant Factor VII
- Limited procedures including blood tests



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What would you do next?



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- Further imaging
- Lymph node biopsy
- Ask a friend
- Search literature
- Give empirical steroids



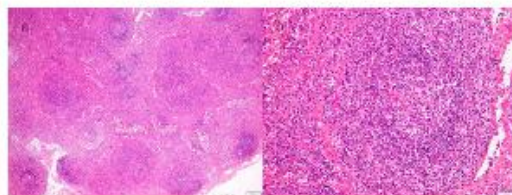


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TAFRO Syndrome: Guidance for Managing Patients Presenting Thrombocytopenia, Anasarca, Fever, Reticulin Fibrosis, Renal Insufficiency, and Organomegaly - MDPI

TAFRO syndrome is an inflammatory disorder of unknown etiology characterized by thrombocytopenia, anasarca, fever, reticulin fibrosis, renal insufficiency, and organomegaly. Despite



Hematology, Transfusion and Cell Therapy

Volume 46, Supplement 4, October 2024, Page S641



ACQUIRED HEMOPHILIA A IN A 3-YEAR-OLD PEDIATRIC PATIENT: A CASE REPORT OF RARE AND POTENTIALLY FATAL BLEEDING DISORDER

Case Reports > [Int J Hematol.](#) 2019 Jun;109(6):737-743. doi: 10.1007/s12185-019-02604-2.

Epub 2019 Jan 24.

Clinicopathological features of TAFRO syndrome complicated by acquired hemophilia A and development of cardiopulmonary arrest that were

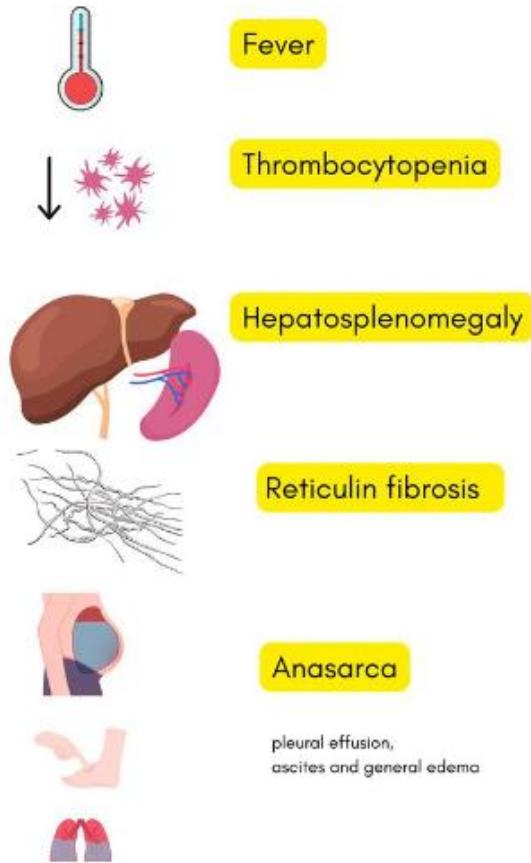




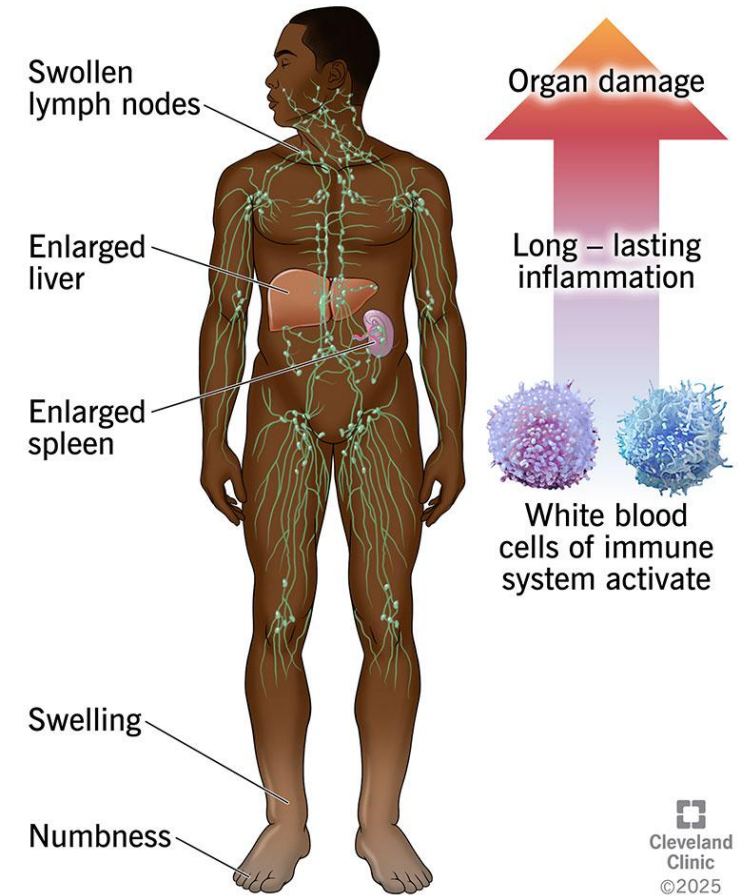
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Case Report- diagnosis

TAFRO Syndrome



Considered a
variant of multi
centric
Castleman
disease





Case Report- diagnosis

- 3/5 clinical criteria
 - Constitutional symptoms, splenomegaly, fluid accumulation
- 6/8 laboratory criteria
 - Elevated CRP, Anaemia, thrombocytopenia, hypoalbuminaemia, renal dysfunction
- Moderate lymphadenopathy in abdomen
 - Not biopsied
- Bone marrow
 - Non-specific but megakaryocyte hyperplasia (no fibrosis)





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Which team should lead her care?



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- Oncology
- Haematology
- Rheumatology
- Renal





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Case report- management

Oral prednisolone (1mg/kg) plus 3 days of
high-dose methylprednisolone (30mg/kg)

Tocilizumab

Rituximab (4 doses at 375mg/m²)

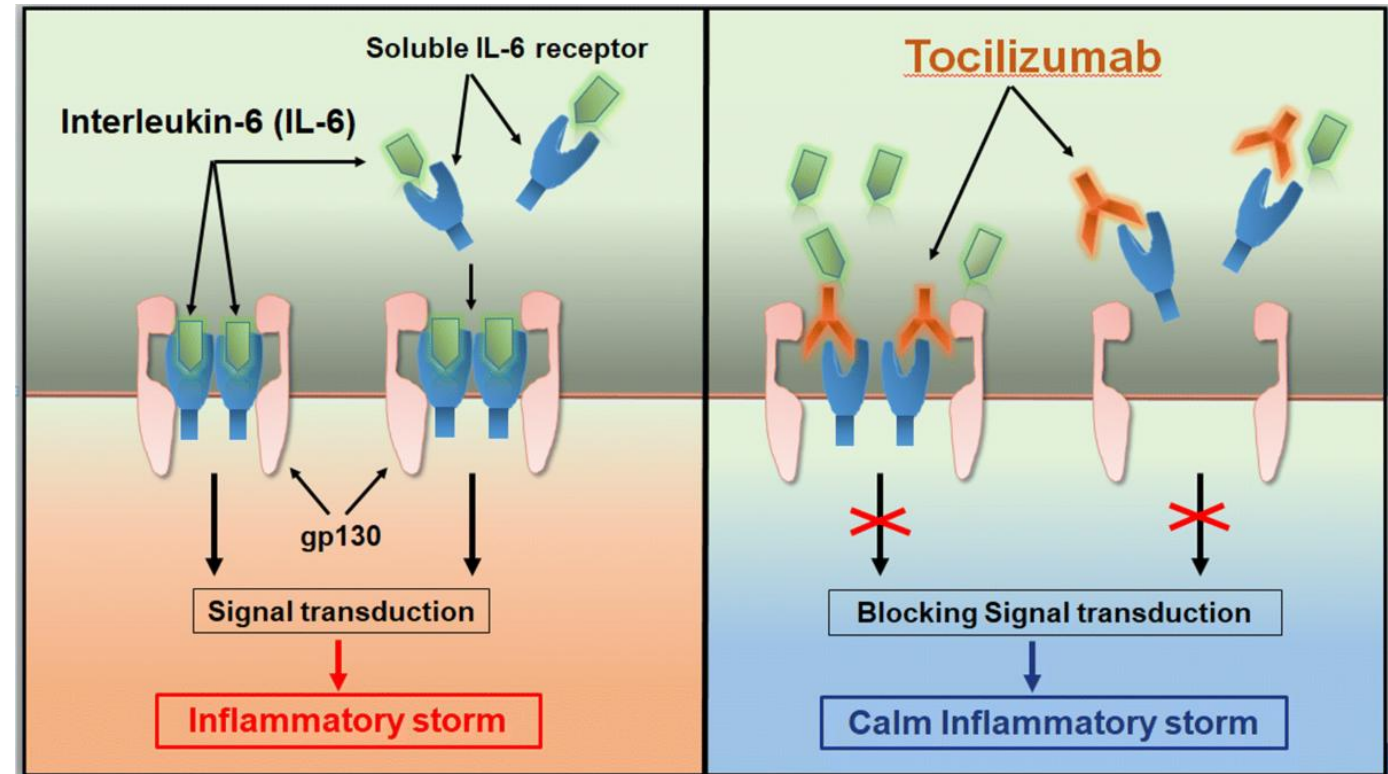
PROGRESS

Renal function normalised within 3 weeks

Resolved haemophilia

Symptoms improved

Remains in remission on fortnightly
tocilizumab





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Discussion





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Take home messages



- Huge benefit to working in large centre with multiple different specialities
- Case reports are not a waste of time
- Use international panels in rare diseases
- Investigate coagulopathy in suspected TAFRO syndrome
- A tissue diagnosis may not be possible in a deteriorating patient

