A RARE CAUSE OF THROMBOCYTOPENIA

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CASE HISTORY

- 41/2 year old, girl, first order child of non consanguineous parents
- Red colored spots over both extremities for 4 days
- No H/O bleeding gums
- No H/O hematemesis or melena
- No H/O drug intake
- No H/O joint pain/ difficulty in breathing
- H/O short febrile illness 1 week ago

CASE HISTORY

- No significant medical illness in past
- Post natal period- uneventful
- Developmental history normal
- Family H/O maternal aunt died of hematological malignancy

EXAMINATION

- Well child
- Multiple petechiae and ecchymotic patches over both upper and lower limbs and neck
- No gum bleeds
- No generalised lymphadenopathy
- No organomegaly

INVESTIGATIONS

- Hb- 9.8 gm/dl
- TC-7,900
- DC-N25L73M2
- **Platelet- 15,000/cu. mm**
- PT- 14/14
- PTT-30/30

PROVISIONAL DIAGNOSIS

ACUTE IMMUNE THROMBOCYTOPENIA

TREATMENT

- Bone marrow aspiration was done
- Started on IV I g

- Bone marrow smear

NO MEGAKARYOCYTES

WHAT ARE WE DEALING WITH?

NOT ITP

AS THERE IS NO MEGAKARYOCYTIC RESPONSE

AMEGAKARYOCYTIC THROMBOCYTOPENIA

(PROBABLY ACQUIRED)

AMEGAKARYOCYTIC THROMBOCYTOPERNIA

- Part of bone marrow failure syndromes
- Thrombocytopenia with absent megakaryocytes in bone marrow
- Associated physical anomalies- absent radius, radio ulnar synostosis
  - Congenital
  - acquired

CONGENITAL AMEGAKARYOCYTIC THROMBOCYTOPENIA

- Presents in the 1st week of life
- Mutation of gene C-MPL – receptor for thrombopoietin
- Serum thrombopoeitin levels are high

ACQUIRED AMEGAKARYOCYTIC THROMBOCYTOPENIA

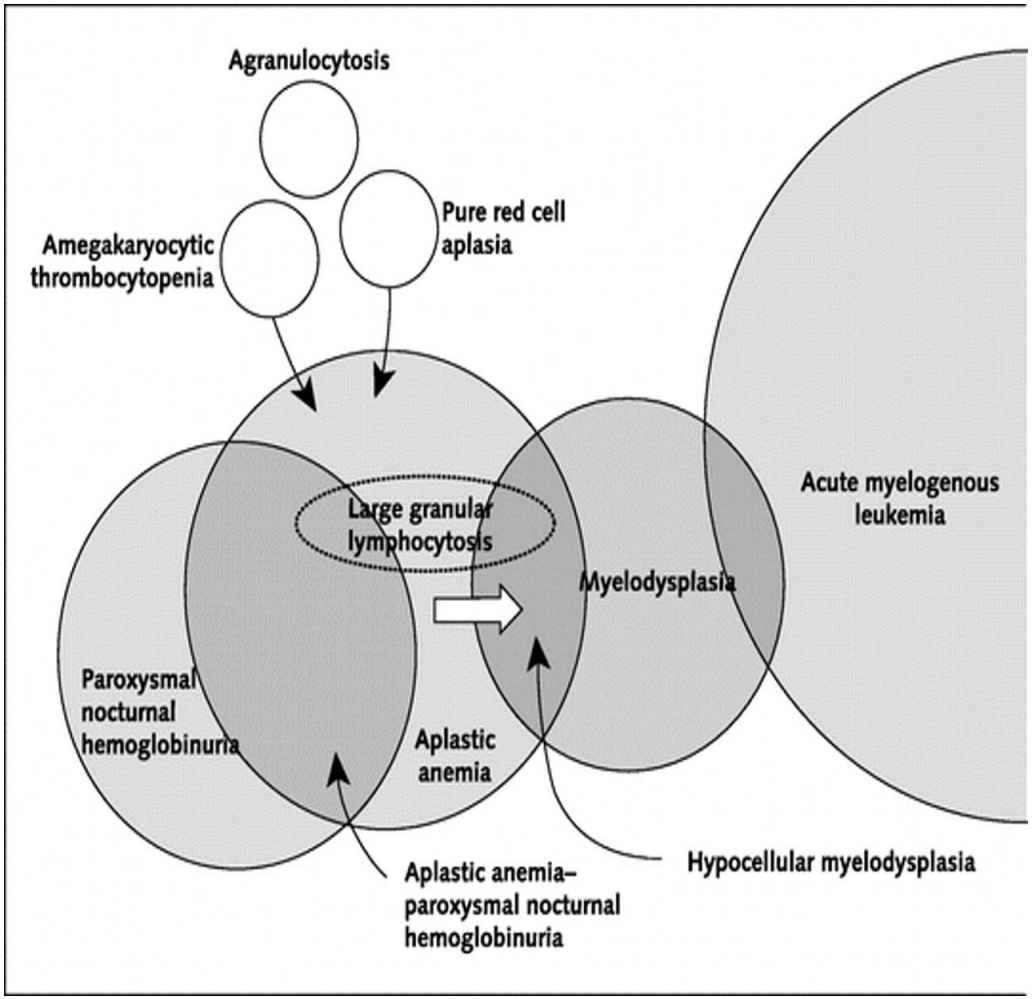
- Viral illness, drugs toxicity, myelophisthic infiltrates of bone marrow
- Antibody and cell mediated immunity- suppression of thrombopoeisis
- Serum thrombopoeitin levels are high
- Usually presents with muco cutaneous bleeds

TREATMENT

- Platelet transfusion
- Regular followup
- Hematopoeitic stem cell transplant

NATURAL HISTORY

- Aplastic pancytopenia
- Myelodysplastic syndrome
- Myeloid leukemia



OUR CHILD

- Repeat platelet count after IVIG 20,000/cu mm
- Started on oral steroids
- Parents counseled about the nature of the disease
- Repeat counts after 2 weeks - pancytopenia Aplastic marrow
- Adviced HSCT

MESSAGE

- Importance of bone marrow smear study in a child with initial presentation like ITP

THANK YOU