LOOK BEYOND THE OBVIOUS

PRESENTO R : DR. VIGNATHA SAJJ A
HISTORY

- Baby X, 5 year old, Girl from Madhya Pradesh

CHIEF COMPLAINTS:
- Fever and body pain x 5 days
- Abdomen pain x 1 day
- One episode of vomiting
HISTORY

HISTORY OF PRESENTING ILLNESS:

- Fever for 5 days – High grade, intermittent, no chills / rigors.
- Headache & pain in the limbs since 5 days
- Abdominal pain since 1 day – Severe
- 1 episode of vomiting – Containing food, not blood/ bile stained
- Tiredness and Breathlessness x 1 day
HISTORY

- No h/o bleeding manifestations (Haematemesis, Melaena)
- No h/o blood transfusions
- No h/o convulsions / Hemiplegia / psychic changes
- No h/o visual disturbances
- No h/o diarrhoea or constipation
- No h/o pica
- No h/o walking barefoot / passing worms in stools
HISTORY

H/o PAST ILLNESS:

- Jaundice one year back - took treatment with the local doctor (Details not known)
- Pain in the limbs off and on since one year of age (Taking pain killers)
- No h/o chronic infections TB, HIV
- No previous h/o treatment with drugs leading to anemia (Chloramphenicol, quinine, sulfonamides)
HISTORY

Antenatal History, Natal History, Post Natal History: Non contributory

B.W t – 2.5 kg, No h/o Neonatal jaundice

DEVELOPMENTAL HISTORY: Normal

IMMUNIZATION / NUTRITIONAL HISTORY: Not contributory
FAMILY AND SIBLING HISTORY:

- No h/o bleeding tendencies, anemia, jaundice, gallstones, blood transfusions, splenectomy in the family.
- 1 year old brother - healthy (according to the parents)
- H/O anemia during pregnancy - mother (no BT)

SOCIO ECONOMIC HISTORY: Class IV
(According to Modified Kuppuswamy scale)
Vital signs
Temperature - 101°F, HR - 126/min
RR - 28/min, BP - 100/60 mmHg
Spo₂ - 96% in Room air

General Physical examination

Pallor + + +. Bony tenderness +

No Icterus / Cyanosis / Clubbing / Koilonychia
/ Lymphadenopathy / Oedema / Dysmorphic facies
/ rashes / signs of avitaminosis
# Examination

## Anthropometry:

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Observed</th>
<th>Expected</th>
<th>Inference (WHO)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weight</td>
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<td>18.2 kg</td>
<td>&lt; 3\text{rd} centile</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Z score &lt; -2</td>
</tr>
<tr>
<td>Weight for Height</td>
<td>12.6 kg</td>
<td>14 kg</td>
<td>15\text{th} centile</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Z score -1</td>
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<tr>
<td>Height</td>
<td>95 cm</td>
<td>109 cm</td>
<td>&lt; 3\text{rd} centile</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Z score &lt; -3</td>
</tr>
<tr>
<td>Mid arm circumference</td>
<td>15 cm</td>
<td>15 – 17 cm</td>
<td>Normal</td>
</tr>
</tbody>
</table>
Abdomen -
Liver - 4 cm below RCM, firm, non tender, rounded margins.
Spleen - 7 cm below LCM, firm, tender, rounded margins.

Cardio Vascular System - Normal.
Other systems - Normal.
LIVER 4 cm below RCM

SPLEEN 7 cm below LCM
Fever with Anaemia and HSM

1. Malaria (Fever, Hepatosplenomegaly, pallor, body pains)

2. Acute leukaemia (Fever, Hepatosplenomegaly, pallor, bone pains)

3. Haemolytic Anaemia (Hepatosplenomegaly, severe pallor, previous history of jaundice)
INVESTIGATIONS

- Hb - 4.4 gm%
- Total count - 3400 cells/dl.
- RBC - 1.58 lakhs
- MCV - 90.3
- MCH - 28.1
- MCHC - 31.1
- Platelets - 54,000

- PT - 14.2/12.2
- PTT - 40.8/28.3
- INR - 1.21
- Reticulocyte count - 3.2
  (Corrected Reticulocyte count - 1.2)
- Blood group - O +ve.
- DCT - negative
INVESTIGATIONS

- Total bilirubin - 3.95
- Direct - 1.13
- SGOT - 65
- SGPT - 38
- Alkaline phosphatase - 109
- LDH - 853
- Total protein - 7.7
- Albumin - 4.0
- Globulin - 3.7
- BUN - 21
- Creatinine - 0.3
- Na⁺ - 139
- K⁺ - 5.2
- Cl⁻ - 102
- HCO₃⁻ - 18
Peripheral smear examination:
- RBC - Microcytic hypochromic with many macrocytes with marked anisopoikilocytes, target cells, fragmented RBC.
- WBC - Leukopenia
- Platelets - Thrombocytopenia, approx 50,000 cells/ mm³
- Hemoparasites - many trophozoites of PLASMODIUM VIVAX seen

Impression: Pancytopenia with plasmodium vivax infection.
INVESTIGATIONS

- Urine routine & culture: Normal
- Blood culture: No growth
TREATMENT GIVEN

- Anti malarials
- Analgesics
- Urgent PRBC transfusion
- Supportive treatment.

Child became afebrile on day 5 of admission and intensity of bone pain decreased
**Fever Chart**

<table>
<thead>
<tr>
<th>O or P.P</th>
<th>9/10</th>
<th>3/10/15</th>
<th>4/10/15</th>
<th>Stools</th>
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<tbody>
<tr>
<td>A.M.</td>
<td>1</td>
<td>2</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td>P.M.</td>
<td>6</td>
<td>10</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>A.M.</td>
<td>6</td>
<td>10</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>P.M.</td>
<td>6</td>
<td>10</td>
<td>2</td>
<td>6</td>
</tr>
</tbody>
</table>

Chloroquine added
Unanswered questions
1. Severe bone pains since one year of age
2. Moderate to massive splenomegaly
3. Prior history of jaundice
**Examination**

- Pallor +
- Liver – Just Palpable.
- Spleen – 4 cm, firm, non tender, round margins.
SIBILING’s investigations:

- Hb - 5.9
- Total counts - 23,000
- Platelet count - 300
- MCV - 96.4
- MCH - 31.6
- MCHC - 32.7
- Reticulocyte count - 2.9
- DCT - Negative
PERIPHERAL SM EAR OF SIBLING SHOWING SICKLE CELLS
Peripheral smear examination:
- RBC - Anisopoikilocytosis, N C N C, sickle cells, target cells, nucleated RBCs seen in plenty. Few microspherocytes seen.
- W BC - corrected count 23,000. Leukocytosis with left shift.
- Platelets - Adequate.
- Hemoparasites - Absent.

Impression: Sickle cell anemia
SICKLING TEST
**CBC:**

<table>
<thead>
<tr>
<th>SNO</th>
<th>TEST NAME</th>
<th>RESULT</th>
<th>UNIT</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>HB</td>
<td>5.9</td>
<td>g/dl</td>
</tr>
<tr>
<td>2</td>
<td>HCT</td>
<td>18.2</td>
<td>%</td>
</tr>
<tr>
<td>3</td>
<td>RBC</td>
<td>1.88</td>
<td>10^6/µL</td>
</tr>
<tr>
<td>4</td>
<td>MCV</td>
<td>96.4</td>
<td>fL</td>
</tr>
<tr>
<td>5</td>
<td>MCH</td>
<td>31.6</td>
<td>pg</td>
</tr>
<tr>
<td>6</td>
<td>MCHC</td>
<td>32.7</td>
<td>g/dl</td>
</tr>
<tr>
<td>7</td>
<td>RDW</td>
<td>27.2</td>
<td>%</td>
</tr>
</tbody>
</table>

**CHROMATOGRAM:**

<table>
<thead>
<tr>
<th>SNO</th>
<th>TEST NAME</th>
<th>RESULT %</th>
<th>REFERENCE RANGE %</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>UNIDENTIFIED PEAK</td>
<td>0.5</td>
<td>&lt;1.0</td>
</tr>
<tr>
<td>2</td>
<td>HAEMOGLOBIN F</td>
<td>22.7</td>
<td>&lt;1</td>
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<tr>
<td>3</td>
<td>PI PEAK</td>
<td>ABSENT</td>
<td>&lt;8</td>
</tr>
<tr>
<td>4</td>
<td>HAEMOGLOBIN A</td>
<td>7.0</td>
<td>94.96</td>
</tr>
<tr>
<td>5</td>
<td>HAEMOGLOBIN A2</td>
<td>1.3</td>
<td>&lt;3.5</td>
</tr>
<tr>
<td>6</td>
<td>HAEMOGLOBIN S</td>
<td>70.8</td>
<td>ABSENT</td>
</tr>
<tr>
<td>7</td>
<td>HAEMOGLOBIN D</td>
<td>ABSENT</td>
<td>ABSENT</td>
</tr>
<tr>
<td>8</td>
<td>HAEMOGLOBIN C</td>
<td>ABSENT</td>
<td>ABSENT</td>
</tr>
<tr>
<td>9</td>
<td>UNIDENTIFIED PEAK</td>
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</tr>
<tr>
<td>10</td>
<td>UNIDENTIFIED PEAK</td>
<td>ABSENT</td>
<td>ABSENT</td>
</tr>
</tbody>
</table>

**Impression:** CHROMATOGRAM SHOWS ELEVATED LEVELS OF HBS 70.8%, HBF 22.7%. PERIPHERAL SMEAR AND CHROMATOGRAM SUGGEST SICKLE CELLS HOMOZYGOUS.

**RBC:** ANISOPOIKILOCYTOSIS, NORMOCRYPIC NORMOCHROMIC, SICKLE CELLS, TARGET CELLS, FRAGMENTED CELLS, NUCLATED RBC'S SEEN IN PLENTY, FEW MICRO - SPIROCYTES SEEN.
PARENTS

Father and mother underwent Hb Electrophoresis
Clinical Pathology

CBC:

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<tr>
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</thead>
<tbody>
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<td>11.6</td>
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<tr>
<td>3</td>
<td>RBC</td>
<td>3.59</td>
<td>10^6/μL</td>
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<tr>
<td>4</td>
<td>MCV</td>
<td>98.3</td>
<td>fl</td>
</tr>
<tr>
<td>5</td>
<td>MCH</td>
<td>32.4</td>
<td>pg</td>
</tr>
<tr>
<td>6</td>
<td>MCHC</td>
<td>33.0</td>
<td>g/dl</td>
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<tr>
<td>7</td>
<td>RDW</td>
<td>15.8</td>
<td>%</td>
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<tbody>
<tr>
<td>1</td>
<td>UNKNOWN UNIDENTIFIED PEAK</td>
<td>ABSENT</td>
<td>&lt;1.0</td>
</tr>
<tr>
<td>2</td>
<td>HAEMOGLOBIN F</td>
<td>&lt;0.8</td>
<td>&lt;1</td>
</tr>
<tr>
<td>3</td>
<td>P3-PEAK</td>
<td>2.8</td>
<td>&lt;8</td>
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<tr>
<td>4</td>
<td>HAEMOGLOBIN A</td>
<td>57.8</td>
<td>94.96</td>
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<td>5</td>
<td>HAEMOGLOBIN A2</td>
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<td>&lt;3.5</td>
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<td>38.5</td>
<td>ABSENT</td>
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<tr>
<td>10</td>
<td>UNKNOWN PEAK</td>
<td>ABSENT</td>
<td>ABSENT</td>
</tr>
</tbody>
</table>

Impression: CHROMATOGRAM SHOWS ELEVATED HBS 38.5%. PERIPHERAL SMEAR AND CHROMATOGRAM ARE IN FAVOUR OF SICKLE CELL TRAIT.

RBC: NORMOCYTIC NORMOCHROMIC
Clinical Pathology

**CBC:**

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<tr>
<td>1</td>
<td>Hb</td>
<td>8.2</td>
<td>g/dl</td>
</tr>
<tr>
<td>2</td>
<td>Hct</td>
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<td>%</td>
</tr>
<tr>
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<td>RBC</td>
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<td>10^6/µL</td>
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<tr>
<td>4</td>
<td>MCV</td>
<td>79.8</td>
<td>fl</td>
</tr>
<tr>
<td>5</td>
<td>MCH</td>
<td>25.5</td>
<td>pg</td>
</tr>
<tr>
<td>6</td>
<td>MCHC</td>
<td>31.9</td>
<td>g/dl</td>
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<tr>
<td>7</td>
<td>RDW</td>
<td>26.4</td>
<td>%</td>
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<tbody>
<tr>
<td>1</td>
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<td>HAEMOGLOBIN F</td>
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<td>&lt;1</td>
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<td>3</td>
<td>P3-PEAK</td>
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<td>UNKNOWN PEAK</td>
<td>0.4</td>
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<tr>
<td>10</td>
<td>UNKNOWN PEAK</td>
<td>ABSENT</td>
<td>ABSENT</td>
</tr>
</tbody>
</table>

Impression: CHROMATOGRAM SHOWS ELEVATED HbS 37.0%. PERIPHERAL SMEAR AND CHROMATOGRAM ARE IN FAVOUR OF SICKLE CELL TRAIT.

**RBC:** NORMOCYTIC NORMOCHROMIC
DIAGNOSIS IN INDEX CASE

PLASMODIUM VIVAX MALARIA
UNDERLYING SICKLE CELL ANEMIA
INFECTION precipitated VASO-OCLUSIV E CRISIS & SPLENIC SEQUESTRATION
Planned treatment:

- **Tab Pencillin prophylaxis**
- **Cap Hydroxyurea**
- **Vaccines against Encapsulated organisms** (Hemophilus.influenza, Streptococcus.Pneumonia, Meningococcus)
SICKLE CELL ANAEMIA

Prevalence in India

- 0 – 18 % in north eastern India
- 0 – 33.5 % in western India
- 22.5 – 44.4 % in central India
- 1 – 40 % in southern India
Sickle Cell Disorders in India

[Map showing distribution of sickle cell disorders in India]
Sickle-Cell Anemia

Normal red blood cell (RBC)

RBCs flow freely within blood vessel

Sticky sickle cells

Sickle cells blocking blood flow

Normal red blood cell section

Normal hemoglobin

Abnormal sickle red blood cell section

Abnormal hemoglobin form strands that cause sickle shape
TREATMENT:

- Hydroxyurea
- Pain management
- Red blood cell transfusion
- Immunization
- Penicillin prophylaxis
- Hematopoietic stem cell transplantation
ANTICIPATORY GUIDANCE

- Spleen palpation.
- Transcranial Doppler ultrasound.
- Screening for pulmonary / renal involvement.
- Retinopathy.
- Echocardiography.
HbAS has protection from *Plasmodium falciparum* and not *Plasmodium vivax*.

HbAS had no effect on the prevalence of symptomless parasitemia.

50% protective against mild clinical malaria.

90% protective against severe or complicated malaria.
AS

P. fal

↑O₂ consumption
↓pO₂

Uptake by macrophages
↓Parasite growth and maturation
↓Adherence to endothelium
Splenic sequestration crises

- Anemia, thrombocytopenia, hypovolemic shock, and sudden death
- Classical age
  - HbSS – 6 months to 3 years
  - HbSβ, HbSC – even in older children (> 10 years)
- Sickle cell anemia (HbSS) – autosplenectomy by 5 years
- Sickle cell disease (HbSβ, HbSC) – splenomegaly can persist in older children
- Treatment – transfusion; later date – splenectomy

## PRECIPITATING FACTORS - CRISIS

| A | Acidosis, anesthetia, anxiety, (high) altitude |
| B | Bouts of infection, bad habits, e.g. smoking, alcohol |
| C | Cold exposure |
| D | Dehydration |
| E | Exercise (vigorous) |
| F | Folate deficiency (e.g. megaloblastic crisis) |
| G | General surgery |
| H | Hypoxia |
| I | Infection |
| O | Other – trauma, menstruation. |
Even when the diagnosis is obvious, seek an explanation for each symptom.


Thank You

It's not my fault that I inherited the sickle-cell gene.

You have to go and check yours too. It will help you to make the right choices when you are older and want to marry, for the sake of your children.