"... Pale and Tired"

GIT Block

PBL; Case 4

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Color Guide:

- **Red:** Relatively important.
- **Black:** Questions.
- **Blue:** Answers (mentioned in case tutorials).
- **Green:** Additional answers/notes.
- **Orange:** Explanation.

Learning Objectives:

On completion of this PBL package, students should be able to:

- Link the histological ultra-structure and biochemical design of red blood cells with their functions.
- Discuss the functions of normal haemoglobin, the types of haemoglobin, and the impact of replacing normal haemoglobin (Hb A) with other types of haemoglobin (such as HbA2, F, or S).
- Discuss the formation of red blood cells and the role of haemopiotic system in their development.
- Discuss the physiology of iron.
- Discuss the mechanisms by which anaemia might occur.
- Apply knowledge from physiology, histology, and pathology to discuss the pathogenesis of Thalassaemia intermediate and beta-Thalassaemia major.
- Use knowledge from basic sciences to interpret symptoms, signs and investigation results of a patient with Beta-Thalassaemia.
- Construct a brief management plan showing management goals, and management options for a patient with Thalassaemia.

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Case Scenario

Key information:

1)Male. 2) 7 years old. 3) Primary school student.

Presenting problems:

1) Always tired.
2) Pallor.
3) Shortness of breath. (After any brief exercise)

Past Medical History:

1- No history of bronchial asthma, blood loss, blood transfusion, or hospital admission.
2- He was born in a small village in **Jeezan**. (+his mother wasn't seen by a doctor during her pregnancy)

Family History: His half-brother is always ill and needs blood transfusion nearly every month. **(it's usually associated with hemolysis).**

Medication and Allergy: Nil.

Social History:

1. He goes to bed early & doesn't complete his homework because of his tiredness.
2. Decreased school performance in most subjects.
3. He is unhappy because he has no friends at his new school.

Clinical Examination:

He looks pale and has no problems with his skeletal muscles.

1- Vital signs: Normal except for tachycardia.

2- Abdominal examination: Liver and spleen are not palpable. (Normal)

3- VS & Respiratory examination: Normal.

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Questions

Before answering the questions below, please read tutorials 1 and 2

Q1: Where do we look for pallor?
Lips, tongue, conjectiva, mucus membranes.
Q2: Why do kidney/liver failure patients look pale?
Patients with **kidney failure** lack erythropoitin hormone → anemia
Patients with **liver failure** lack protien synthesis → low hemoglobin synthesis → anemia
Q3: Why do anemic patients usually present with tachycardia?
Tachycardia serves as a compensatory mechanism.

Q4: How does anemia impact school performance?
Due to ↓ hemoglobin **(↓ oxygenation)** → ↓Ability to concentrate.

Q5: What are some possible causes of microcytic hypochromic anemia?
The main cause is iron deficiency secondary to other conditions such as malabsorption or thalassemia.

Q6: What's the main difference between alpha thalassemia and beta thalassemia blood films?
Presence of **nucleated red blood cells** in Beta thalassemia major.

Q7: Why does the blood film of thalassemic patients show hypochromasia?
Due to ↓ Hemoglobin.

Q8: Why does the blood film of thalassemic patients show polychromasia?
Because as the hemolysis inceases, the bone marrow tries to compensate for the loss by increasing its rate of production of RBCs which also produces immature RBCs known as "reticulocytes". The bluish color of reticulocytes mixes along with the red color of RBC resulting in "polychromasia".

Q9: What are target cells?
Also known as Codocytes, or Mexican hat cells, are red blood cells that have the appearance of a shooting target with a bullseye.

Q10: Why is iron supplementation contraindicated in patients with thalassemia?
Because these patients already suffer from **iron overload**.
Imbalance in proteins controlling iron intake (such as **hepcidin**) → ↑ iron absorption → iron overload

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Q11: What hemoglobin level would you request in order to find out if your patient is alpha or beta thalassemic?
Hemoglobin A2
If it was less than 1.5 → Alpha thalassemia.
If it was more than 3.5 → Beta thalassemia.

Q12: In what way can the hemoglobin electrophoresis test help?
It enables us to differentiate between types/percentages of each hemoglobin present in patient's blood (normal/abnormal) and therefore helps in making more accurate diagnosis.

Q13: Why do patients with beta thalassemia major need blood transfusion?
Because their hemoglobin level is **severely decreased** (around 1.0-5.9 g/dl). So, a blood transfusion will help maintain hemoglobin level in their blood.

Q14: Why don't patients with beta thalassemia minor need blood transfusion?
Because their hemoglobin level would probably be in the range of 7.5-8.5 g/dl (**not severely decreased**) so we wouldn't put our patient at risk of iron toxicity *-secondary to blood transfusion-* just to correct this mild change.

Q15: What are some certain occasions that require patients with beta thalassemia minor to transfuse blood?
1. Presence of severe infection.
Infection → ↑ metabolism (and fever) → ↑ oxygen requirement.
2. Hyperactive spleen.
Rapid destruction of RBCs → severe ↓ Hemoglobin.

Q16: How can we differentiate between beta thalassemia major, intermedia or minor? By the hemoglobin level, blood changes and **the onset** of signs and symptoms.

Q17: Why was the Hemoglobin F level increased in this case?
Because when beta chains decrease in number, gamma proteins are produced in compensation. This results in an increase in hemoglobin F. (Which is formed of 2 alpha, 2 gamma)

Q18: How to differentiate iron deficiency anemia from beta thalassemia?
1. Serum ferritin level
2. Mentzer index.

Q19: How to distinguish hypochromasia from hyperchromasia?
By mean cell hemoglobin. (MCH)
Q20: What does an increase in red blood cell distribution width (RDW or RCDW) indicate?
High RDW values indicate greater variation in size. (**Anisocytosis**)

General notes

- Hemoglobin is a substance present normally in the red blood cells, it transports the oxygen and canbone dioxide between the lungs and the tissues.
- Beta thalassemia is a hereditary defect in the make up of hemoglobin so it will not be able to function normally.
- There are three forms of beta thalassemia: Thalassemia minor, thalassemia intermedia, and thalassemia major. (which is also called **Cooley's anemia**)

Hemoglobin electrophoresis for Beta thalassemia:

Thalassemia major:
Hb A= 0
Hb A2= 1.0-5.9
Hb F= > 94
With free alpha chains
Thalassemia intermediate:
Hb A= present
Hb A2= 5.4-10
Hb F= 30-73
Thalassemia minor:
Hb A= > 90
Hb A2= 3.5-8
Hb F= 1-2

Good Luck!