

HAEMATOLOGICAL DISORDERS

Introduction

Blood is an extremely complex fluid, composed of both formed elements (red cells, white cells, platelets) and plasma. RBCs (erythrocytes) are the most common formed elements, carrying Oxygen and haemoglobin.

- White blood cells function as mediators of immune responses to infection or other stimuli of inflammation.
- Platelets are the formed elements that participate in coagulation. Plasma is largely water, electrolytes, and plasma proteins. The plasma proteins most important in blood clotting are the coagulation factors.
- A group of haematological disorders characterized by any disturbance in the physiological and morphological changes in the blood cell is called haematological disorder.

Clinical consideration— Most common haematological disorder is Anaemia.

Classification of Anaemia.

1. Morphological classification.
 - a. Normocytic normochromic anaemia.
 - b. Macrocytic normochromic anaemia
 - c. Macrocytic hypochromic anaemia.
 - d. Microcytic hypochromic anaemia.
2. Etiological classification.
 - a. Haemorrhagic anaemia.
 - b. Haemolytic anaemia.
 - c. Aplastic anaemia.
 - d. Anaemia due to other disease.
 - e. Nutritional deficiency anaemia.
 - Iron deficiency anaemia.
 - Protein deficiency anaemia.
 - Vitamin B12 anaemia.
 - Folic acid anaemia.

Iron deficiency anaemia

Definition.

- Iron deficiency anaemia is most common nutritional disorder in the world. It develops due to inadequate availability of iron for haemoglobin synthesis. RBCs are microcytic and hypochromic. Dietary deficiency of iron is most common cause in the developing country.
- Blood loss in this case may result from relatively benign disorders, such as peptic ulcer, arteriovenous malformations, or angiodysplasia (small vascular abnormalities along the intestinal walls). More serious causes are inflammatory bowel disease and malignancy.
- Daily requirement of iron is about 10 to 20 mg. It varies person to person. About 80% of the functional iron is found in haemoglobin, myoglobin and iron-containing enzymes.

Etiopathogenesis.

- Dietary deficiency of iron is most common cause in the developing country. Menstrual blood loss, premenopausal is also major cause in the female.
- Helicobacter pylori infection, partial gastrectomy, bleeding disorders and haemoptysis are also the cause of iron deficiency anaemia.
- Hepcidin (25-amino acid peptide) produced by liver, is also cause for the anaemia because it exports the iron to plasma after recycling of senescent erythrocyte via macrophages phagocytosis and lysis.
- Hepcidin binds to ferroportin, a transmembrane protein, inducing its internalization and lysosomal degradation. When iron stores are low, hepcidin production is reduced and ferroportin molecules are expressed on the basolateral membrane of enterocytes, where they transfer iron from the cytoplasm of enterocytes to plasma transferrin. Conversely, when iron stores are adequate or elevated, hepcidin production is increased, resulting in the internalization of ferroportin and reduced export of iron into plasma.

Clinical manifestations.

- Increase in the BMR (basal metabolic rate).
- Heart disease occurs (Tachycardia, angina, heart attack).
- Spleen enlargement, liver enlargement.
- Hyperventilation and dyspnoea.
- Menorrhagia, oligomenorrhoea/amenorrhoea.
- Dysphagia, anorexia, nausea vomiting.
- Headache and dizziness.

- Fatigue and weakness.
- Skin and mucosa turn yellow in colour.
- Brittle hair and nails, atrophy of papilla in tongue.

Pharmacological managements—

- Oral iron— ferrous sulphate, ferrous aminoate, ferrous gluconate, ferrous succinate, carbonyl iron, iron calcium complex.
- Parenteral iron— Iron sucrose, iron dextran, iron isomaltoside, ferric carboxy maltose, ferric pyrophosphate citrate.

Non-pharmacological management—

- Daily intake of iron richest sources are green vegetables such as lettuce, spinach, asparagus, and broccoli. Certain fruits (e.g., lemons, bananas, melons) and animal sources (e.g., liver, meat especially dark meat).
- Digestive activity is very important factor for the absorption of iron, so follow all the regular regimen including yoga, exercise, balance diet etc.
- During the pregnancy/menstrual flow iron deficiency is common, so more iron rich supplement and more take care needed.

Megaloblastic anaemia

Definition.

- Megaloblastic anaemia is developing due to the deficiency of maturation factor called folic acid. In this condition, RBCs are not matured and DNA synthesis is also defective/impairment leads to ineffective haematopoiesis and distinctive morphologic changes, including abnormally large erythroid precursors and red cells. The RBCs are megaloblastic and hypochromic.
- Decreased intake can result from either a nutritionally inadequate diet or impairment of intestinal absorption. Megaloblastic anaemia that results from a deficiency of folic acid is identical to that encountered in vitamin B12 deficiency. Thus, the diagnosis of folate deficiency can be made only by demonstration of decreased folate levels in the serum or red cells.

Etiopathogenesis—

The three major causes of folic acid deficiency are Decreased intake of iron, Increased requirements of iron, Impaired utilization of iron.

- Humans are entirely dependent on dietary sources for their folic acid requirement, which is 50 to 200 µg daily. The richest sources are green vegetables such as lettuce, spinach, asparagus, and broccoli. Certain fruits (e.g., lemons, bananas, melons) and animal sources (e.g., liver). The folic acid in these foods is largely in the form of polyglutamates. Intestinal conjugases split the polyglutamates into monoglutamates that are readily absorbed in the proximal jejunum. During intestinal absorption they are modified to 5-methyltetrahydrofolate, the normal transport form of folate. The body's reserves of folate are relatively modest, and a deficiency can arise within weeks to months if intake is inadequate.
- The megaloblastic anaemia is often accompanied by general malnutrition and manifestations of other avitaminoses, including cheilosis, glossitis, and dermatitis. Malabsorption syndromes, such as sprue, can lead to inadequate absorption of this nutrient, as can diffuse infiltrative diseases of the small intestine (e.g., lymphoma).

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Pharmacological managements—Iron containing preparation is given as-

- Oral iron— ferrous sulphate, ferrous aminoate, ferrous gluconate, ferrous succinate, carbonyl iron, iron calcium complex.
- Parenteral iron— Iron sucrose, iron dextran, iron isomaltoside, ferric carboxy maltose, ferric pyrophosphate citrate.
- Maturation factors— Hydroxocobalamin, methyl cobalamin, cyanocobalamin, folic acid/leucovorin.

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