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Practical Standard Prescriber



ABM Abdullah

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4

CHAPTER

Hematology

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ANEMIA

It is a clinical condition characterized by reduced hemoglobin concentration according to the age and sex of the individual.

Classification:

- Etiological (based on cause)
- Morphological [based on morphology of red blood cell (RBC)].

Etiological:

- *Hemorrhagic anemia (due to blood loss)*:
  - Acute: Trauma, postpartum bleeding, hematemesis, melena and epistaxis
  - Chronic: Hookworm, hemorrhoid, excessive menstrual loss, and bleeding peptic ulcer.
- *Deficiency anemia*:
  - Iron deficiency anemia
  - Vitamin B12 and folic acid deficiency (megaloblastic anemia).
- *Dyshemopoietic anemia (less production of RBC)*:
  - Aplastic anemia (bone marrow failure, may be primary or secondary)
  - Anemia of chronic disorder: Systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), sideroblastic anemia, chronic kidney disease (CKD), hypothyroidism and malignancy.
- *Hemolytic anemia (due to breakdown of RBC)*:
  - Genetic: Red cell membrane defect (e.g. hereditary spherocytosis, elliptocytosis and stomatocytosis), hemoglobin abnormality (thalassemia, and sickle cell anemia), and enzyme defects [glucose-6-phosphate dehydrogenase (G6PD) deficiency and pyruvate kinase deficiency]
  - Acquired: Autoimmune, toxic, mechanical and infectious causes.

Morphological [depending on mean corpuscular volume (MCV) and mean corpuscular hemoglobin concentration (MCHC)]:

- Normocytic normochromic anemia (normal MCV and MCHC)
- Microcytic hypochromic anemia (low MCV <76 fL, low MCHC <30 g/dL)
- Macrocytic anemia (high MCV >96 fL)

- Dimorphic anemia (combination of two cell lines—macrocytes and microcytes).

Causes of normocytic normochromic anemia: Hemorrhagic and hemolytic anemia, aplastic anemia, anemia of chronic disorder, malignancy, endocrine disease, and sideroblastic anemia.

Causes of microcytic hypochromic anemia: Iron deficiency, thalassemia, sideroblastic and anemia of chronic disorder.

Causes of macrocytic anemia:

- *Macrocytosis with megaloblastic marrow*: Vitamin B12 or folic acid deficiency
- *Macrocytosis with normoblastic marrow*: Chronic liver disease (CLD), chronic alcoholism, hypothyroidism, hemorrhage, hemolysis, sideroblastic anemia, pure red cell aplasia, and azathioprine.

DIMORPHIC ANEMIA

When both microcytes and macrocytes are found in peripheral blood film (PBF), this is called dimorphic anemia.

Causes: Combined iron, B12 or folate deficiency, and sideroblastic anemia, during treatment of anemia.

Signs that may point to a specific cause of anemia:

- *Hemolytic anemia*: Triad of anemia, jaundice and splenomegaly
- *Iron deficiency anemia*: Angular stomatitis, cheilitis, glossitis and koilonychia
- *Glossitis*: Iron deficiency anemia, vitamin B12 deficiency, and folate deficiency
- *Vitamin B12 deficiency (megaloblastic anemia)*: Neurological changes (dementia, optic atrophy, and features of subacute combined degeneration of spinal cord) and lemon yellow tint.

Investigations of anemia: Detailed history, physical examination, and relevant laboratory investigations are done.

History:

- Dietary history and malabsorption
- History of bleeding (hemorrhoid, epistaxis, hematemesis, melena and menorrhagia in female)
- *In female*: Multiple pregnancies, repeated abortion, and menorrhagia
- *Drug history*: Nonsteroidal anti-inflammatory drugs (NSAIDs) drugs causing bone marrow suppression (e.g. cytotoxic drugs), and drugs causing hemolysis (e.g. sulfasalazine and methyldopa)
- *History of surgery*: Gastrectomy or partial gastrectomy and ileal surgery (causes deficiency of vitamin B12)
- Family history (in hereditary hemolytic anemia)
- History of chronic disease (e.g. SLE, RA, CKD and CLD).

Clinical examination: See above.

Laboratory investigations:

- Complete blood count (CBC), erythrocyte sedimentation rate (ESR), platelet, and PBF (to see normocytic, microcytic, macrocytic or dimorphic)
- Reticulocyte count—high in hemolytic anemia
- MCV and MCHC

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- Bone marrow examination—to detect megaloblastic anemia, aplastic anemia, bone marrow infiltration (secondary deposit), and ring sideroblasts (in sideroblastic anemia)
- Other investigations according to suspicion of cause.

Investigations of microcytic hypochromic anemia (low MCV and low MCHC):

- *For iron deficiency anemia*: Serum iron, total iron-binding capacity (TIBC), and serum ferritin
- *For hereditary hemolytic anemia*: Hemoglobin electrophoresis and skeletal survey
- *For sideroblastic anemia*: According to history, bone marrow examination (to see ring sideroblasts)
- *For anemia of chronic disease*: Test should be done according to the cause.

Investigations for macrocytic anemia (high MCV): Serum B12 and folic acid assay. Bone marrow study, if needed.

SIDEROBLASTIC ANEMIA

It is defined as a group of anemia characterized by refractory anemia, variable number of hypochromic cells in the peripheral blood, excess iron, and ring sideroblast.

Types:

- Hereditary
- *Acquired, which may be primary and secondary*:
  - Primary sideroblastic anemia (refractory anemia with ring sideroblast)
  - Secondary sideroblastic anemia may be due to—(1) Drugs and chemicals [isonicotinylhydrazide (INH), alcohol and lead], (2) Hematological disease [myelofibrosis, polycythemia rubra vera (PRV), myeloma, Hodgkin's lymphoma, hemolytic anemia, and leukemia], (3) Inflammatory disease (RA, and SLE), and (4) Others (carcinoma, myxedema and malabsorption).

Treatment: Treatment of primary cause, e.g. withdrawal of drug and stop alcohol. Some cases may respond to pyridoxine and folic acid. Correction of anemia is done by blood transfusion.

IRON DEFICIENCY ANEMIA

Anemia secondary is due to deficiency of iron.

Causes:

- *Bleeding due to any cause*: Gastrointestinal tract (GIT) (hemorrhoid, colorectal carcinoma, carcinoma of stomach, diverticulitis and angiodysplasia of colon), and menorrhagia in female
- Hookworm (also schistosomiasis)
- Less intake of food
- Malabsorption
- More demand (e.g. pregnancy).

Symptoms: Weakness, vertigo, dizziness, lightheadedness, tingling, numbness, anorexia, weight loss, palpitation, breathlessness on exertion, and pica (eating of unusual material, e.g. earth).

Signs: The patient is pale and anemic. Koilonychia, glossitis and angular stomatitis.

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Investigations:

- *Test to confirm iron deficiency anemia*:
  - Complete blood count with PBF (microcytic hypochromic blood picture)
  - Serum iron, TIBC and ferritin (low iron, low ferritin, and increased TIBC).
- *Test to find out causes*:
  - Stool for ova or cyst of hookworm and occult blood test
  - Upper gastrointestinal (GI) endoscopy (to see esophageal varices, peptic ulcer, and carcinoma stomach)
  - Proctoscopy (hemorrhoid), sigmoidoscopy or colonoscopy (neoplasm, polyp, diverticulum, ulcer and angiodysplasia of colon)
  - Ultrasonography (USG) of abdomen (any mass and fibroid uterus in female).

Treatment:

- Severe anemia should be corrected by blood transfusion
- *Iron therapy*: Oral ferrous sulfate, ferrous gluconate or ferrous fumarate, for 3–6 months after hemoglobin is normal to replenish the iron store. If the patient is unable to take iron orally, it can be given intravenous (IV)
- Treatment of cause (e.g. menorrhagia and hemorrhoid).

MEGALOBLASTIC ANEMIA

It is a type of anemia which is due to deficiency of vitamin B12, folic acid or both, characterized by macrocyte in peripheral blood and megaloblast in bone marrow.

Causes of megaloblastic anemia due to vitamin B12 deficiency:

- *Diet*: Strict vegetarians
- *Stomach pathology*: Total or partial gastrectomy, carcinoma of stomach, and pernicious anemia
- *Small bowel pathology*:
  - Ileal disease: Crohn's disease and ileal resection
  - Pancreatic exocrine insufficiency: Vitamin B12 deficiency occurs in 30% patients
  - Motility disorders: It can cause bacterial overgrowth and vitamin B12 deficiency
  - Fish tapeworm (Diphyllobothrium latum).

Pernicious Anemia

It is an autoimmune disease in which there is atrophy of gastric mucosa with loss of parietal cells causing intrinsic factor deficiency. So, vitamin B12 is not absorbed due to deficiency of intrinsic factor. There are anti-intrinsic factor antibodies and also anti-parietal cell antibodies in serum.

Symptoms of megaloblastic anemia: Weakness, malaise, weight loss, breathlessness and sore mouth. Paresthesia, poor memory, depression, hallucinations, and visual disturbance.

Signs:

- Angular cheilitis. Tongue—smooth and shiny with atrophy of papillae
- Skin pigmentation, lemon yellow tint, and vitiligo

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- *Neurological*:
  - Peripheral neuropathy
  - Spinal cord (subacute combined degeneration): Loss of vibration and position sense (posterior column lesion), and upper motor neuron signs (corticospinal tract lesion)
  - Cerebral: Dementia
  - Optic atrophy.

Investigations:

- *CBC*: Low hemoglobin (Hb)% may indicate pancytopenia. PBF shows macrocytes and hypersegmented neutrophils
- Serum vitamin B12 (low)
- Parietal cell and intrinsic factor antibodies for pernicious anemia
- *Bone marrow aspiration*: Shows increase cellularity and megaloblasts
- *Others*: Low reticulocyte count and high lactic acid dehydrogenase (LDH).

Treatment: Blood transfusion in severe anemia. Injection hydroxycobalamin 1,000 µg intramuscular (IM) daily six dose, for 2 or 3 days apart. Then 1,000 µg every 3 months (lifelong for pernicious anemia).

Causes of megaloblastic anemia due to folic acid deficiency:

- Less intake of vegetables
- *Malabsorption*: Due to any cause (e.g. celiac disease and tropical sprue)
- *Increased demand*: Pregnancy and hemolysis
- *Drugs*: Methotrexate and phenytoin.

Investigation: Serum folate level (low) and red cell folate level (low).

Treatment: Tablet folic acid 5 mg is given daily for 3 weeks and then it is given 5 mg weekly. Prophylactic folic acid should be given in pregnancy and also in hemolytic anemia.

Note: Folic acid alone should not be given if there is vitamin B12 deficiency, otherwise it may aggravate neurological lesion. In such cases, both should be given.

HEMOLYTIC ANEMIA

Anemia occurs due to destruction or breakdown of RBC.

There are two types:

- *Inherited*:
  - Red cell membrane abnormality: Hereditary spherocytosis and elliptocytosis
  - Red cell enzyme deficiency: G6PD deficiency and pyruvate kinase deficiency
  - Hemoglobinopathy: Thalassemia, sickle cell disease, and other hemoglobinopathies.
- *Acquired*:
  - Autoimmune:
    - Warm antibodies (primary or idiopathic and secondary in SLE, RA, chronic lymphatic leukemia (CLL), and lymphoma, drugs such as methyldopa and quinidine)
    - Cold antibodies (primary or idiopathic, secondary in Mycoplasma pneumoniae, and lymphoma).

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- Nonimmune:
  - Mechanical (microangiopathic hemolytic anemia and prosthetic valve)
  - Infection (falciparum malaria)
  - *Chemicals*: Dapsone, sulfasalazine and maloprim
  - *Acquired membrane abnormality*: Paroxysmal nocturnal hemoglobinuria (PNH).

Evidence of Hemolysis

- *Clinical*: Triad of anemia, jaundice and splenomegaly (in hereditary hemolytic anemia)
- *In the blood*:
  - High serum bilirubin
  - High urinary urobilinogen
  - Low haptoglobin
  - High LDH
  - High reticulocytes
  - Erythroid hyperplasia in bone marrow.

THALASSEMIA

It is an inherited disorder characterized by impaired hemoglobin production due to partial or complete failure to synthesize specific type of globin chain.

There are two types:

1. *Beta-Thalassemia*: Less production of β chain, causing less production of HbA. It is of two types—
   i. β-Thalassemia major: HbA is less and HbF is high
   ii. β-Thalassemia minor: HbA2 is high.
2. *Alpha-Thalassemia*: Inadequate production of α chain, so less HbA, HbF and HbA2, as all of them contain α chain.

Beta-thalassemia Major

It is characterized by reduced or absent synthesis of β chain of globin. Family history may be present.

Symptoms: Weakness, palpitation and dizziness. Yellow coloration of eyes, high colored urine. Feeling of a mass in the left upper abdomen (due to splenomegaly).

Note: If the patient develops severe abdominal pain, likely cause is cholelithiasis. There may also be splenic infarction and acute pancreatitis.

Signs: Triad of hemolytic anemia (anemia, jaundice and splenomegaly). Frontal and parietal bossing, and mongoloid facies with prominent malar bones. Short stature and retardation of growth.

Investigations:

- CBC and PBF (shows microcytic hypochromic blood picture), and reticulocyte count (high)
- Hemoglobin electrophoresis
- *Others*: X-ray of skull and hand, and other skeletal survey, and serum bilirubin (high). Serum iron profile—ferritin (high), iron and TIBC.

Treatment:

- *Correction of anemia*: Blood transfusion to keep Hb% above 10 g/dL, every 4 months (because life span of RBC is 4 months)

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- Folic acid 5 mg daily, to be continued
- Iron-containing drugs and diet are avoided (iron can only be given if deficiency)
- Repeated blood transfusion may cause hemosiderosis, which can be prevented by chelating agent. (see below)
- Injection erythropoietin
- Hydroxyurea 1–2 g daily may be helpful
- *Specific therapy*: Allogeneic bone marrow transplantation. Also, gene therapy
- Splenectomy may be needed in some cases
- Genetic counselling should be offered.

Complications of Repeated Blood Transfusion

- Hemosiderosis (usually when more than 30–50 L of blood is transfused)
- Infections such as hepatitis B, C and D, and human immunodeficiency virus (HIV).

Prevention of Hemosiderosis

It can be prevented by using chelating agent desferrioxamine (1.5–2 g with each unit of blood), usually given subcutaneously in the anterior abdominal wall with infusion pump for 12 hours. It may also be given with infusion drip (normal saline or aqua). Oral chelating agents such as deferiprone and deferasirox. Vitamin C 200 mg daily orally also helps in iron excretion.

Thalassemia Minor

Usually asymptomatic or mild anemia.

Investigations: CBC shows microcytic hypochromic blood picture. Hemoglobin electrophoresis shows high A2.

Differential diagnoses: It confuses with iron deficiency anemia. However, anemia is more marked in iron deficiency and relatively less in thalassemia minor. Also, in iron deficiency, there is low iron, low ferritin, and high TIBC.

Alpha-Thalassemia

It is characterized by reduced or absent alpha globin chain synthesis. Normally, adults have four alpha globin chain genes.

- When 1 alpha gene is absent—patient is hematologically normal (silent carrier)
- When 2 alpha genes are absent—called alpha thalassemia trait. The patient is clinically normal, life expectancy is normal
- When 3 alpha genes are absent—the patient has HbH disease. There is chronic hemolytic anemia with variable severity. Examination shows severe anemia with splenomegaly
- If all 4 alpha globin chains are absent—affected fetus is stillborn due to hydrops fetalis.

AUTOIMMUNE HEMOLYTIC ANEMIA

It is group of hemolytic anemia that occurs due to red cell autoantibodies causing increased red cell destruction. Antibodies may be IgG or IgM. Antibodies are of two types depending on temperature at which the antibody reacts with red cells.

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- Warm antibodies bind at 37°C, common (80% cases). Antibody is IgG
- Cold antibodies bind at 4°C but can bind up to 37°C in some cases. Antibody is IgM. Less common (20% cases).

Warm Autoimmune Hemolytic Anemia

Occurs at all ages, common in middle age, more in females.

Causes:

- Primary or idiopathic (50% cases)
- *Secondary*: Infection (*M. pneumoniae*, infectious mononucleosis, and cytomegalovirus), collagen diseases (SLE and RA), drugs (methyldopa and quinidine), neoplastic (CLL, and lymphoma), and others (carcinoma and sarcoidosis).

Clinical features: May be asymptomatic or features of primary cause or features of hemolytic anemia (anemia, jaundice and splenomegaly).

Investigations: CBC with PBF shows macrocytes and spherocytes. Direct Coombs or antiglobulin test is positive. Serum IgG is high.

Treatment: Underlying cause should be treated, offending drugs must be stopped. Prednisolone 1 mg/kg orally. When Hb and reticulocyte count are normal, dose can be reduced slowly over 10 weeks. Blood transfusion is done in case of severe anemia. If there is no response to prednisolone, splenectomy should be considered. If splenectomy is not possible, azathioprine or cyclophosphamide may be given. Anti-CD20 monoclonal antibody, rituximab is helpful in some cases.

Cold Agglutinin Disease

It is an autoimmune hemolytic anemia which is due to cold antibodies. It is of two types—(1) Cold hemagglutinin disease (CHAD), and (2) paroxysmal cold hemoglobinuria (PCH).

Chronic cold agglutinin disease occurs in elderly and may be associated with an underlying low-grade B-cell lymphoma. It causes low-grade intravascular hemolysis with cold, painful, blue fingers, toes and ears or nose (called acrocyanosis).

Causes:

- Idiopathic in most cases
- *Secondary*: Lymphoma, CLL, SLE and Waldenström's macroglobulinemia. Also in *M. pneumoniae* and infectious mononucleosis.

Investigation: The CBC with PBF shows cold agglutination at low temperature. Direct Coombs test is positive with complements. Serum IgM is high.

Treatment: Treatment of underlying causes. If idiopathic—patients must keep extremities warm, especially in winter. Some respond to steroid; blood transfusion may be needed in severe anemia. Folic acid should be given.

SICKLE CELL ANEMIA

It is a hereditary disorder, inherited as autosomal recessive trait, in which the red cells contain an abnormal hemoglobin called HbS.

There are two types:

1. Homozygotes produce abnormal β-chains that make hemoglobin S (called AS). This causes clinical syndrome of sickle cell disease

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2. Heterozygotes produce normal and abnormal β-chains that make normal HbA and HbS. This causes sickle cell trait, which is clinically asymptomatic.

When hemoglobin S is deoxygenated and red cell membrane is distorted, then it produces sickle-shaped cells.

Clinical features: Usually present in childhood before 2 years.

- Features of hemolytic anemia, frequent infection, leg ulcers, gall stone, and dactylitis (due to microinfarction of medulla of carpal and metatarsal bones)
- Splenomegaly is present, but repeated infarction of spleen causes atrophy (autosplenectomy) at the age of 8 years. So spleen is no longer palpable
- *May present with following crises called sickle cell crises*:
  - Vaso-occlusive crisis: Plugging of small vessels in bone produces acute severe bone pain. Mesenteric infarction causes acute abdominal pain. Papillary necrosis may occur in kidney. Cerebral infarction is due to involvement of brain
  - Sickle chest syndrome: Bone marrow infarction results in fat emboli to lungs, which causes further infarction, leading to ventilatory failure
  - Sequestration crisis: There is splenic pooling of red cells and hypovolemia, leading to circulatory collapse and death. Liver sequestration can also occur
  - Aplastic crisis: Red cell aplasia causes very low hemoglobin.

Investigations:

- Complete blood count with PBF shows sickle cells, target cells, and features of hyposplenism
- Reticulocyte count—high
- Sickle solubility test
- Hemoglobin electrophoresis shows high HbS, absence of HbA, and 2–20% HbF.

Treatment:

- Phenoxymethylpenicillin is given to protect pneumococcal infection
- Folic acid. Blood transfusion, if severe anemia. Hydroxyurea may be helpful
- Vaccination against *Pneumococcus*, *Meningococcus*, *Haemophilus influenzae* B, hepatitis B, and influenza
- Allogeneic stem cell transplantation from human leukocyte antigen (HLA)-matched siblings may be curative.

MICROANGIOPATHIC HEMOLYTIC ANEMIA

It is a type of mechanical hemolytic anemia in which anemia is associated with fragmented RBC (schistocyte and helmet cell). Red cell fragmentation is due to contact between red cells and abnormal intima of partially thrombosed, narrowed or necrotic vessels.

Causes:

- Disseminated intravascular coagulation (DIC)
- Thrombotic thrombocytopenic purpura (TTP)
- Hemolytic uremic syndrome (HUS)
- Disseminated malignancy
- Vasculitis

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- Metallic cardiac valve prosthesis
- Malignant hypertension.

Investigations: CBC with PBF (shows anemia with fragmented RBC). Others—bilirubin and high reticulocyte count.

Treatment: Treatment of primary cause.

PAROXYSMAL NOCTURNAL HEMOGLOBINURIA

It is an acquired clonal abnormality of red cells, which are destroyed by activated complement resulting in intravascular hemolysis. Platelet and granulocyte are involved, causing thrombocytopenia and leukopenia.

Clinical features: Commonly, urine, voided at night and in morning on waking from sleep, is dark colored. Hemolysis may be precipitated by infection, surgery or iron therapy.

Complications:

- Venous thrombosis involves mesenteric, portal (Budd–Chiari syndrome) or cerebral veins, and calf muscles
- Aplastic anemia may precede PNH in 25% cases, acute myeloid leukemia (AML) may occur
- *Others*: Infection, iron deficiency, and pigment gallstone.

Investigations:

- Complete blood count shows anemia, leukopenia and thrombocytopenia
- Ham acid serum test may be positive
- Flow cytometric analysis of red cells with CD55 and CD59 has replaced Ham test
- Bone marrow may be hypoplastic or aplastic.

Treatment:

- Supportive (blood transfusion, and iron), and prednisolone (in some cases)
- *In marrow failure*: Antithymocyte globulin and cyclosporin or bone marrow transplantation
- Recombinant humanized monoclonal antibody (eculizumab) may be helpful
- *Antiplatelet may be used*: Aspirin and clopidogrel
- Gene therapy.

Prognosis: Survival is 10–15 years.

HEMOLYTIC UREMIC SYNDROME

It is characterized by rapid onset of microangiopathic hemolytic anemia, thrombocytopenia, and acute renal failure (triad) due to thrombosis in small arteries and arteriole.

Causes: Verotoxin producing organisms, such as enterohemorrhagic *Escherichia coli* 0157:H7, also *E. coli* 0104:H4. HUS may be diarrheal (D+HUS) or non-diarrheal HUS (D-HUS or atypical HUS).

Clinical features: Common in children, <3 years. It usually follows gastroenteritis.

- Fever, vomiting and diarrhea, often bloody, called diarrhea associated HUS (D+HUS)

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- Intravascular hemolysis is followed by oliguria or anuria
- Purpura, anemia, bleeding, drowsiness and hypertension may occur.

Investigations:

- *Complete blood count*: Anemia and marked thrombocytopenia. PBF shows features of microangiopathic hemolytic anemia (schistocyte) and spherocyte
- *Creatinine*: Very high
- *Other features of intravascular hemolysis*: High bilirubin, high LDH, reduced haptoglobin, and increased reticulocyte count
- Confused with DIC, but coagulation tests are normal in HUS.

Treatment: In diarrheal HUS, treatment is mainly supportive—

- Correction of water and electrolyte balance, and maintenance of nutrition
- Control of hypertension
- FFP transfusion and dialysis may be needed
- Heparin or antiplatelet drug may be helpful
- Plasmapheresis is not helpful.

In atypical HUS:

- Supportive, as above
- Plasmapheresis or plasma transfusion
- *Monoclonal antibody (anti-CD5 antibody)*: Eculizumab may be effective.

Prognosis: About 5% die in acute episode, 5% develop CKD, and 30% develop persistent proteinuria.

THROMBOTIC THROMBOCYTOPENIC PURPURA

Thrombotic thrombocytopenic purpura (TTP) is a disorder of unknown etiology characterized by fever, microangiopathic hemolytic anemia, thrombocytopenia, neurological signs, and renal failure.

Clinical features: Common in young female. Fever and purpuric spot due to thrombocytopenia, fluctuating neurological features, microangiopathic hemolytic anemia, and renal failure.

It confuses with HUS, but neurological features are absent in HUS. These two conditions are probably the part of same disorder.

Diagnosis: Diagnostic *pentad* in TTP—

1. Fever
2. Neurological features: Headache, seizure and coma
3. Thrombocytopenia
4. Microangiopathic hemolytic anemia
5. Renal failure.

Investigations:

- *Complete blood count and PBF*: Thrombocytopenia and fragmented RBC (schistocyte). Reticulocytes (high)
- *Bleeding time*: Prolonged
- *Bilirubin and LDH*: High. Also haptoglobin is reduced
- *Coagulation screen*: Activated partial thromboplastin time (APTT), D-dimer, fibrin degradation product (FDP), and prothrombin time (PT) are all normal. Usually no DIC.

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Treatment:
- Plasma exchange. Fresh frozen plasma or cryoprecipitate infusion
- Pulse intravenous (IV) methylprednisolone may be given
- Rituximab is also very helpful
- Platelet transfusion is contraindicated, as it may aggravate the disease.

Prognosis: Mortality 90% in untreated and 10–30% in treated cases.

APLASTIC ANEMIA

It is a disorder due to bone marrow failure leading to pancytopenia. Diagnosis is done by bone marrow aspiration and biopsy.

Causes: It may be primary and acquired or secondary.
- *Primary*: Cause unknown
- *Acquired or secondary*:
  - Drugs: Cytotoxic drugs, antibiotics (chloramphenicol and sulfonamide), antithyroid drugs (carbimazole), carbamazepine, and penicillamine
  - Secondary deposits in bone marrow
  - Radiotherapy
  - Chemicals: Benzene, lindane and dichlorodiphenyltrichloroethane (DDT)
  - Infection: Viral hepatitis, Epstein-Barr virus (EBV) infection, HIV and parvovirus infection
  - Others: Lymphoma, multiple myeloma, and PNH.

Symptoms (Primary):
- *Features of anemia*: Weakness, palpitation, dizziness and vertigo
- *Bleeding manifestations*: Gum bleeding, nasal bleeding, multiple bleeding spots such as bruise, and ecchymosis in different parts of the body
- *Infection*: Sore throat, other infections, and septicemia.

Signs:
- Anemia. Bleeding spots (e.g. purpura, ecchymosis and bruise)
- No lymphadenopathy, hepatomegaly and splenomegaly or bony tenderness (which may be present in secondary deposit or secondary causes of aplastic anemia).

Note: In secondary aplastic anemia, features of primary are the predominant features.

Investigations:
- *Complete blood count*: Shows pancytopenia (anemia, leukopenia and thrombocytopenia)
- *Bone marrow (aspiration and trephine biopsy)*: Dry tap or acellular or hypocellular marrow.

Treatment:
- Removal of causative agent, if any
- *Supportive therapy*: Blood transfusion and control of infection
- *Allogeneic bone marrow transplantation (with HLA-matched siblings)*: If the patient is below 30 years with severe idiopathic aplastic anemia
- *In older patient or if HLA-matched donor is not available*: Immunosuppressive therapy with cyclosporine and antithymocyte globulin should be given. Tacrolimus may be used

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Key Features

- The book *Practical Standard Prescriber* presents in brief the etiology, clinical presentation, investigations and management of various diseases encountered in clinical practice on a day-to-day basis
- Covers diseases pertaining to almost all the body systems with emphasis on genetics, poisoning, psychiatric disorders, electrolyte and acid-base imbalance, and specific diseases pertaining to vitamin deficiency
- Presented in a format which is easy to understand with accentuated bullet points for quick referral
- Written by an expert well known for his academic inclination and capability
- The book is a ready reckoner for general practitioners, residents, and postgraduate students.

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