Haematology

Blood transfusion reaction

a) Febrile reaction

Diagnostic tools

1. Patient complaint of fever, chills & rigor (If isolated temperature ≥38°C, or rise of 1–2°C from baseline).

2. Patient otherwise normal.

Management

1. Stop the transfusion temporarily.

2. Paracetamol tablet or suppository.

3. Restart the transfusion at a slower rate.

4. Observe the patient more frequently.

b) Urticaria

Mild allergic reaction

1. Stop the transfusion.

2. Give chlorpheniramine 10 mg IV slowly.

3. Restart the transfusion at a slower rate.

4. Observe frequently.

c) Severe allergic reaction

Diagnostic tools

1. Bronchospasm (SOB), abdominal pain.

2. Angioedema, hypotension.

Management

1. Discontinue the transfusion permanently.

2. Start O2 inhalation.

3. Nebulize with salbutamol.

4. Give chlorpheniramine 10 mg IV slowly.

5. If severe bronchospasm or hypotension then give inj. adrenaline 0.5 mg IM stat.

6. Send clotted blood sample to the transfusion laboratory.

7. Take down blood unit & giving set & return intact to blood bank with all other used/unused unit.

d) ABO incompatibility

Management

1. Take down blood unit & giving set & return intact to blood bank with all other used/ unused unit

2. Start intravenous normal saline infusion

3. Monitor urine output- maintain urine output >100ml/ hour. Give frusemide if urine output fall

4. Treat DIC with appropriate blood products

5. Inform hospital transfusion department immediately.

Iron deficiency anaemia

Diagnostic tools

1. Patient complaint of weakness, palpitation, vertigo.

2. History of menorrhagia, melaena, haematemesis may be present.

3. Investigation to confirm iron deficiency anaemia - PBF - microcytic, hypochromic anaemia (MCV and MCHC reduced), s. ferritin-low.

4. Investigation to determine cause of iron deficiency anaemia-stool for parasites, endoscopy of upper GIT, colonoscopy etc.

Management

1. Diet- iron containing foods like banana, lal sak, kochu sak etc.

2. Ferrous sulphate 200 mg 8 hourly for 3-6 months. If patient intolerant to ferrous sulphate 8 hourly dose then reduce dose to 12 hourly or ferrous gluconate 300 mg 12 hourly.

3. Blood transfusion-indication

a) If patient have angina

b) Heart failure

c) Evidence of cerebral anoxia

4. Parentral iron-indication

i) Malabsorption

ii) Chronic gut disease

iii) Inability to tolerate any oral preparation

5. Treatment of the underlying cause like treatment of menorrhagia, treatment of piles, treatment of PUD etc.

Follow up:

1. Hb%- Hb should increase 1 gm/dl/week

2. Reticulocyte count- reticulocytosis is evident after 7-10 days.

N:B: usually capsule formulations dissolve in ileum but iron absorption take place at stomach and duodenum, so iron capsule has little or no benefit. Iron should not be prescribe with PPI, because acidic media is neccessary to absorb iron.

Megaloblastic anaemia

Diagnostic tools

1. Symptoms of anaemia.

2. Patient may be vegetarian.

3. May have history of gastrectomy or ileal resection.

4. Investigation-PBF-macrocytic anaemia (MCV-increased commonly > 120 fL), serum vit-B12 low or red cell folate level low.

Management

1. Diet- normal

2. Inj. hydroxycobalamine 1000 µg IM 6 doses 2-3 days apart then 3 monthly lifelong; if cause of megaloblastic anaemia cannot be removed. In the presence of neurological involvement, a dose of 1000 µg on alternate days until there is no further improvement, followed by maintenance as above.

3. Folic acid 5 mg daily for 3 week, then 5 mg weekly for lifelong; if cause of megaloblastic anaemia cannot be removed.

4. Ferrous sulphate 200 mg 8 hourly, if initial responses is not maintained & PBF is dimorphic (both microcytic and macrocytic).

5. Blood transfusion-if severe angina or heart failure.

Splenectomized patient

Splenectomy usually done in traumatic injury in spleen and in thallassemia, due to development of hypersplenism.

Management

1. Vaccinate the patient with (2-3 weeks before elective surgery. If emergency surgery is done then vaccination should be done after surgery.

-P Pneumococcal vaccine

-P H. influenza type B vaccine

-P Meningococcus C vaccine

-P Influenza vaccine

2. Pneumococcal reimmunization should be given every 5 yearly.

3. Influenza reimmunization should be given yearly basis.

4. Penicillin V 500 mg 12 hourly lifelong.

5. Should carry bracelet or card.

6. If patient develop any fever then prompt action should be taken, blood, urine should be cultured, because these patient rapidly develop septicaemia.

7. In sepsis, patients should be resuscitated and given IV antibiotics to cover pneumococcus, Haemophilus and meningococcus, according to local resistance patterns.

8. The risk of cerebral malaria is increased in the event of infection.

9. Animal bites should be promptly treated with local disinfection and antibiotics, to prevent serious soft tissue infection and sepsis.

Autoimmune haemolytic anaemia

Diagnostic tools

1. Symptoms of anaemia.

2. Patient may complaint of yellow coloration of eye and urine.

3. Features of underlying cause of autoimmune hemolytic anaemia may predominat (SLE).

4. Investigation-PBF-spherocytes and polychromasia on the blood film. The diagnosis is confirmed by the direct Coombs or antiglobulin test.

Management

1. 1st line treatment-steroid -prednisolone 1 mg/kg/day orally, when Hb% normalized, reticulocytosis resolved; dose of steroid should be reduced over 10 weeks.

2. 2nd line treatment-if the haemolysis fails to respond to glucocorticoids or can only be stabilized by large doses. These include immunomodulation/suppression and splenectomy. If splenectomy is not appropriate, alternative immunosuppressive therapy with rituximab (choice of drug), azathioprine, ciclosporin, mycophenolate or cyclophosphamide may be considered.

3. Blood transfusion- when developed heart failure or unabated fails in Hb%.

Paroxysmal nocturnal haemoglobinuria (PNH)

Diagnostic tools

1. Patient complaint -red brown color urine particularly in morning.

2. Urine R/E- haemoglobinuria.

Management:

1. Diet- normal.

2. Blood transfusion if anaemia.

3. Folic acid supplementation

4. Treatment of thrombosis.

5. Recently anticomplement C5 monoclonal Ab ecluzimab is used.

Sickle cell anemia

Management

1. Folic acid supplementation

2. Penicillin V 500 mg BD (to protect against pneumococcal infection which may be lethal in the presence of hyposplenism).

3. Hydroxycarbamide.

4. Vaccinate against pneumococcus, H. influenza & Hepatitis B.

Treatment of vaso-occlusive crisis

1. O2 inhalation.

2. Rehydration with normal saline.

3. Antibiotic.

4. Analgesia with opioids.

5. Exchange transfusion

* In life threatening crisis

* Or to prepare the patient for surgery.

Chronic myeloid leukaemia

Diagnostic tools

1. Patient present with the complaint of lump/discomfort in left side of the abdomen, weakness, loss of appetite.

2. On examination- huge splenomegaly.

3. Investigation

a) CBC with PBF-WBC count increased (10 thousands to 6 lacs), PBF-predominant cells are neutrophils and myelocytes. Myeloblasts usually constitute less than 10% of all white cells. There is often an absolute increase in eosinophils and basophils, and nucleated red cells are common.

b) Bone marrow examination to confirm the diagnosis and phase of disease.

c) S. uric acid-usually increased due to increase cell breakdown.

N:B: CML has 3 phases; chronic phase, accelerated phase and blast crisis phase. Before management identification of phase of CML is mandatory. Progression of the disease from chronic phase is characterized by i) increase in blast cell count ii) reduction of platelet count iii) increase in basophil count.

Management

Chronic phase

1. First line therapy- tyrosine kinase inhibitors (TKIs) like imatinib, nilotinib and dasatinib.

If fail to response or progress on therapy- use another TKI.

2. Hydroxyurea- is still useful in palliative situations.

3. Interferon is used in women planning pregnancy.

Accelerated or blast crisis phase

1. Accelarated phase-for patients in accelerated phase, TKI therapy is indicated, most commonly with nilotinib or dasatinib.

2. Blast crisis phase-the type of blast cell should be determined. Second or third generation TKIs such as dasatinib are used in combination with chemotherapy to try and achieve remission. In younger and fitter patients an allogeneic HSCT.

3. Hydroxycarbamide can be an effective single agent and low-dose cytarabine can also be used palliatively in older patients.

For better treatment CML patients should be referred to haematologist.

Chronic lymphatic leukaemia

Diagnostic tools

1. The onset is usually insidious. Around 70% of patients, the diagnosis is made incidentally on a routine CBC. Presenting problems may be anaemia, infection, painless lymphadenopathy, and systemic symptoms such as night sweats or weight loss.

2. CBC with PBF-mature lymphocytosis (>5 × 109/L) with characteristic morphology and cell surface markers.

3. Bone marrow-is not essential for the diagnosis of CLL, but may be helpful in difficult cases and to see prognosis

Staging and management

Stage A- no anaemia or thrombocytopenia and fewer than three areas of lymphoid enlargement. No treatment is required. Treatment is required only if there is evidence of bone marrow failure, massive or progressive lymphadenopathy or splenomegaly, systemic symptoms such as weight loss or night sweats, a rapidly increasing lymphocyte count, autoimmune haemolytic anaemia or thrombocytopenia.

Stage B- no anaemia or thrombocytopenia, with three or more involved areas of lymphoid enlargement. Treatment is necessary.

Stage C- anaemia and/or thrombocytopenia, regardless of the number of areas of lymphoid enlargement. Treatment is necessary. For better treatment CLL patients should be referred to haematologist.

Supportive management

1. Corticosteroid- in bone marrow failure or autoimmune cytopenias.

2. Blood transfusion if anaemia or thrombocytopenia.

3. Antibiotic - if infection.

4. Radiotherapy-if lymph node causing discomfort or local obstruction & symptomatic splenomegaly.

5. Splenectomy- to decrease autoimmune destruction of RBC, hypersplenism & to relieve massive splenomegaly.

Myelodysplastic syndrome

Management

1. Blood transfusion

2. Platelet transfusion.

3. Erythropoietin & GCSF (granulocyte colonoy stimulating factor).

4. Allogenic bone marrow transplantation in younger patient.

For better treatment MDS patients should be referred to haematologist.

Multiple myeloma

Diagnostic tools

1. Patient usually old aged (60-70 years)-presented with generalized bodyache, headache (due to hyper viscosity)

2. Investigation-for diagnosis any of the two criteria will require

    i) Increase malignant plasma cells in the bone marrow.

    ii) Serum and/or urinary paraprotein present

    iii) Skeletal lytic lesions present (usually on skull x-ray).

Management

Asymptomatic - if patients are asymptomatic with no evidence of end-organ damage (e.g. kidneys, bone marrow or bone), treatment may not be required. Patient should be monitored closely for the development of end-organ damage.

Symptomatic patient

Immediate supportive management

1. Plenty of fluid intake.

2. Analgesia to relieve pain.

3. Bisphosphonates (for hypercalcaemia and to delay other skeletal related events).

4. Allopurinol (to prevent urate nephropathy).

5. Plasmapharesis-in hyperviscosity.

Chemotherapy with or without HSCT (haemopoietic stem cell transplantation)

For first-line therapy in older patients, thalidomide combined with the alkylating agent melphalan and prednisolone (MPT). Lenalidomide is approved first-line treatment for patients not eligible for transplantation and who are intolerant of, or unsuitable for, thalidomide.

In younger, fitter patients, standard treatment includes firstline therapies, such as cyclophosphamide, thalidomide and dexamethasone (CTD) or bortezomib (velcade), thalidomide and dexamethasone (VTD) to maximum response, and then autologous HSCT. In all patients who have achieved maximal response, lenalidomide maintenance has been shown to prolong the response.

When myeloma progresses, treatment is given to induce a further plateau phase. Proteosome inhibitor bortezomib and lenalidomide have been used as second- and third-line therapy, as appropriate. A second-generation proteasome inhibitor, carfilzomib, and the anti-CD38 antibody daratumumab show promise in relapsed/ refractory disease. Responding patients may benefit from a second autologous HSCT.

Radiotherapy- localized bone pain not responding to simple analgesic & pathological fracture, emergency treatment of spinal cord compression complicating extradural plasmacytomas.

Aplastic anaemia

Diagnostic tools

1. Patient present with the symptoms of anaemia (weakness), leucopenia (infection-fever) and thrombocytopenia (bleeding manifestations-gum bleeding, epstaxis, menorrhagia etc).

2. On examination-ESR high, anaemia present, bruise, purpura, echymoses may be present but no bony tenderness, no lymphadenopathy, no hepatosplenomegaly.

3. Investigation-PBF normocytic normochromic anaemia, bone marrow-hypocellular.

Management

1. Diet- normal.

2. Antibiotic for infection.

3. Blood transfusion to correct anaemia.

4. Immune suppressive therapy for older patient with-ciclosporin & antithymocyte globulin.

5. Patients under 35 years of age with severe idiopathic aplastic anaemia -allogeneic HSCT.

Myelofibrosis

Management

1. Folic acid 5 mg daily

2. Red cell transfusions for anaemia

3. Hydroxycarbamide may help control spleen size, the white cell count or systemic symptoms.

4. Splenectomy- if symptomatic pancytopenia, enlarged spleen with features of hypersplenism.

5. HSCT may be considered for younger patients.

6. Ruxolitinib (an inhibitor of JAK-2) is effective at reducing systemic symptoms and splenomegaly.

Polycythaemia rubra vera

Diagnostic tools

1. Patients over the age of 40 years and presents either as an incidental finding of a high haemoglobin, or with symptoms of hyperviscosity, such as lassitude, loss of concentration, headache, dizziness, blackouts, pruritus and epistaxis. Some patients present with manifestations of peripheral arterial disease or a cerebrovascular accident.

2. On examination- patients are often plethoric and the majority have a palpable spleen at diagnosis.

3. Investigation- Hb-adult female >16.5 gram/dl or haematocrit > 0.48; adult males 18.0 g/dl or haematocrit > 0.52.

Management

1. Aspirin 75 mg daily (reduces the risk of thrombosis).

2. Venesection 400-500 ml every 5-7 days until PCV <45%.

3. Hydroxycarbamide or interferon-alfa (suppression of marrow proliferation).

4. Radioactive phosphorous for older patient.

ITP (idiopathic thrombocytopenic purpura)

Diagnostic tools

1. Patient usually female, presented with the complaint of bleeding manifestation like gum bleeding, purpura, easy bruising or sometimes epistaxis or menorrhagia. (fever usually absent).

2. On examination-anaemia and splenomegaly present.

3. Investigation-PBF-usually normal except platelet count reduced, bone marrow- increase in megakaryocytes.

Management

1. Stable compensated ITP & platelet count >30 thousands do not require treatment, except at times of increased bleeding risk, such as surgery and biopsy.

2. First line therapy for patient with spontaneous bleeding

   A. Prednisolone 1 mg/ kg daily or dexamethasone (40 mg daily for 4 days.

   B. If response to steroid is slow & severe haemostatic failure-prednisolone & IVIg.

   C. Persistent or potentially life threatening bleeding should be treated with platelet transfusion in addition to other therapies.

3. Relapse should be treated with reintroduction of steroid.

4. Second line therapy- If a patient has two relapses or primary refractory disease. The options for second-line therapy include the thrombopoietin receptor agonists (TPO-RA) eltrombopag and romiplostim, splenectomy and immunosuppression.

5. If significant bleeding persists despite splenectomy- low dose steroid, immunosuppressive therapy with rituximab, ciclosporin & tacrolimus.

Haemophilia

Diagnostic tools

1. Patient male, present with prolonged bleeding in case of any cut injury, sponteneous bleeding into skin, muscle and joints.

2. Investigation-serum factor VIII and IX reduced.

Management

Haemophillia A

1. Resting of the bleeding site by bed rest or splint.

2. In severe hemophilia A intravenous infusion of factor VIII concentration

3. In mild to moderate hemophilia A vasopressin receptor agonist DDAVP 0.3 µg/ kg IV or subcutaneously. Alternatively same effect can be achieved by intranasal administration of 300 µg.

4. Prophylaxis - factor VIII can be administered 2 or 3 times per week.

Haemophillia B

Factor IX concentration intravenous infusion.

Acute leukaemia

Diagnostic tool

1. Fever usually high grade.

2. Generalized weakness.

3. Bleeding manifestations e.g. - gum bleeding, epistaxis, etc.

4. On examination - anaemia, generalized lymphadenopathy, bony tenderness, hepato-splenomegaly.

5. Investigation-CBC with PBF-Hb%-reduced, ESR- high, WBC count-increased; PBF-increase blast cell. Bone marrow examination-to confirm diagnosis.

Management (Supportive)

1. Blood transfusion (before BT PBF should be done).

2. Antibiotic to control infection.

3. Tab. Paracetamol (500mg) 1+1+1

For specific management patient should be referred to Haematologist.

Haemolytic anaemia

Diagnostic tool

1. Generalized weakness

2. No fever, bleeding manifestation

3. On examination- anaemia present, no lymphadenopathy, no bony tenderness but hepatosplenomegaly present.

4. Investigation-Hb%-reduced, PBF-microcytic hypochromic anaemia, target cell present, Hb electrophoresis- confirm the diagnosis.

Management

1. Blood transfusion to correct anaemia.

2. Folison 5 mg daily

3. Splenectomy in moderate to severe haemolysis with complications (anaemia and gallstones).

Dietary advice:

1) AvqiY wmivc Lv‡eb bv |

2) Lvevi †jvnvi cv‡Î ivbœv Ki‡eb bv |

3) Lvevi cv‡Î ev ciciB dj LvIqv hv‡e bv |

4) AwaK †jŠnhy³ Lvevi Gwo‡q Pjyb | †hgb-Miy I Lvwmi gvsm, KwjRv, wW‡gi Kzmyg, Bwjk gvQ, ˆK gvQ, wPZj, †Usiv, †QvU gv‡Qi ïUwK| KPz kvK, jvj kvK, cvjs, cyuB kvK, dzj Kwc, cyw`bv I a‡b cvZv, wkg, eiewU, gUikywU, KvK‡ivj, KuvPv †cu‡c, mRbv | Avbvim †e‡`bv, kwidv, †LRyi, ZigyR

Kb©‡d·, †jŠn ‡gkv‡bv wkï Lv`¨- wb‡Wv, †mwijvK †Qvjv I †Qvjvi Wvj ¸o, ev`vg, wKmwgm, wZj, cvb, wRiv, a‡b, mwilv |

5) Kg †jŠnhy³ Lvevi Lv‡eb, †hgb- KvZjv gvQ, †evqvj gvQ, gv¸o gvQ, cywU gvQ evav Kwc, wgwó Avjy, Kijv, kmv, wS½v, KuvPvKjv, jvD, PvjKzgov| wjPz, cvKv †cu‡c, Kgjv †jey, †ej, Rvgiæj, AvgjwK, KvMwR †jey|

Pvj, gq`v| gmyi| gay, `ya, `y»RvZ Lv`¨- দই, Qvbv BZ¨vw` |

An approach to diagnosis of anaemia from examination findings:

* Anaemia + bleeding manifestation + no lymphadenopathy + no bony tenderness + no hepatosplenomegally - Aplastic anaemia.

* Anaemia + no bleeding manifestation + no lymphadenopathy + no bony tenderness + hepatosplenomegaly - Haemolytic anaemia.

* Anaemia + bleeding manifestation + lymphadenopathy + bony tenderness + hepatosplenomegaly - Acute leukaemia.