

MCQs & Cases (20 Questions)

1. A 58-year-old man presents with headache, pruritus after a hot shower, and splenomegaly. Labs: Hb 19 g/dL, Hct 58%. Erythropoietin is low. Which mutation is most likely?

- A) BCR-ABL
- B) JAK2
- C) MPL
- D) CALR

2. The most common complication of untreated Polycythemia Vera is:

- A) Deep vein thrombosis
- B) Hemorrhage
- C) Renal failure
- D) Pancytopenia

3. Which of the following laboratory findings differentiates secondary polycythemia from Polycythemia Vera?

- A) Low oxygen saturation
- B) Presence of JAK2 mutation
- C) Splenomegaly
- D) Increased uric acid

4. A 45-year-old athlete lives at high altitude. Hb: 18 g/dL, Hct: 56%, EPO: high. Diagnosis?

- A) Polycythemia Vera
- B) Secondary polycythemia
- C) Hemoconcentration
- D) Essential thrombocythemia

5. In Polycythemia Vera, which lab result would you expect?

- A) Low hematocrit**
 - B) Elevated erythropoietin**
 - C) Decreased erythropoietin**
 - D) Normal WBC and platelets**
-

1. → B) JAK2

🚩 95–99% of PV patients have JAK2 mutation.

2. → A) Deep vein thrombosis

🚩 Hyperviscosity + abnormal platelets → thrombosis is #1 complication.

3. → A) Low oxygen saturation

🚩 Secondary polycythemia = ↑ EPO + hypoxia (altitude, lung disease).

4. → B) Secondary polycythemia

🚩 Athlete at high altitude → ↑ RBC due to hypoxia.

5. → C) Decreased erythropoietin

🚩 PV suppresses EPO via negative feedback.

6. A 67-year-old woman with Polycythemia Vera develops sudden chest pain. ECG shows ST-elevation MI. What is the underlying mechanism?

- A) Hypocalcemia**
 - B) Hyperviscosity with thrombosis**
 - C) Autoimmune destruction of RBCs**
 - D) Elevated bilirubin**
-

7. Which symptom is classically associated with Polycythemia Vera?

- A) Night sweats**
 - B) Itching after hot shower**
 - C) Petechial rash**
 - D) Gum bleeding**
-

8. Which of the following is NOT a feature of Polycythemia Vera?

- A) Splenomegaly**
 - B) Elevated RBC mass**
 - C) Elevated erythropoietin**
 - D) Hyperviscosity**
-

9. Treatment of choice for symptomatic Polycythemia Vera:

- A) Hydroxyurea + phlebotomy**
- B) Chemotherapy**
- C) Bone marrow transplant**
- D) Corticosteroids**

10. Which of the following best explains the pruritus in PV?

- A) Increased histamine release from basophils
- B) Hyperbilirubinemia
- C) Elevated bile salts
- D) Iron overload

6. → B) Hyperviscosity with thrombosis
✚ Thrombosis is main cause of MI/stroke in PV.

7. → B) Itching after hot shower
✚ Due to histamine release from basophils.

8. → C) Elevated erythropoietin
✚ PV has low EPO, not elevated.

9. → A) Hydroxyurea + phlebotomy
✚ Standard treatment to reduce viscosity & control cell counts.

10. → A) Increased histamine release from basophils
✚ Explains aquagenic pruritus.

11. A 62-year-old man with PV presents with sudden left-sided weakness. CT brain shows ischemic stroke. What is the most likely cause?

- A) JAK2 mutation
- B) Thrombosis due to high platelets
- C) Low oxygen saturation
- D) Vitamin B12 deficiency

12. A 70-year-old man presents with fatigue, hepatosplenomegaly, and "tear-drop" RBCs on blood smear. Bone marrow biopsy: fibrosis. Diagnosis?

- A) Polycythemia Vera
- B) Essential thrombocythemia
- C) Primary Myelofibrosis
- D) Aplastic anemia

13. Which of the following lab results is most consistent with Primary Myelofibrosis?

- A) Pancytopenia
 - B) Elevated EPO
 - C) Isolated thrombocytosis
 - D) Low uric acid
-

14. A 60-year-old man with renal cell carcinoma develops polycythemia. Which lab result would you expect?

- A) Low erythropoietin**
 - B) High erythropoietin**
 - C) JAK2 mutation positive**
 - D) Splenomegaly**
-

15. In PV, which type of mutation is most common?

- A) Tumor suppressor loss**
 - B) Tyrosine kinase activation**
 - C) DNA repair mutation**
 - D) p53 mutation**
-

11. → B) Thrombosis due to high platelets

🚩 Stroke in PV → thrombosis is the mechanism.

12. → C) Primary Myelofibrosis

🚩 Tear-drop cells + BM fibrosis = hallmark.

13. → A) Pancytopenia

🚩 Fibrosis crowds out marrow → ↓ RBCs, WBCs, platelets.

14. → B) High erythropoietin

🚩 RCC secretes EPO → secondary polycythemia.

15. → B) Tyrosine kinase activation

🚩 JAK2 mutation → constitutive TK signaling → uncontrolled growth.

16. Which condition is considered a myeloproliferative neoplasm (MPN) along with PV?

- A) Chronic lymphocytic leukemia**
 - B) Essential thrombocythemia**
 - C) Hodgkin's lymphoma**
 - D) Multiple myeloma**
-

17. Which laboratory marker suggests high cell turnover in PV?

- A) Elevated LDH and uric acid**
 - B) Elevated bilirubin only**
 - C) Low alkaline phosphatase**
 - D) Low serum ferritin**
-

18. Best first-line management of hyperviscosity in PV:

- A) Aspirin**
- B) Phlebotomy**
- C) IV fluids**
- D) Bone marrow transplant**

19. A patient with long-standing PV develops anemia, splenomegaly, and bone marrow fibrosis. This is called:

- A) Blast crisis**
 - B) Spent phase**
 - C) Myelodysplastic syndrome**
 - D) Hemolytic anemia**
-

20. Which is the most feared long-term complication of Polycythemia Vera?

- A) Transformation to acute leukemia**
- B) Vitamin B12 deficiency**
- C) Pulmonary embolism**
- D) Liver cirrhosis**

16. → B) Essential thrombocythemia

📌 MPN group includes: PV, ET, PMF, CML.

17. → A) Elevated LDH and uric acid

📌 High turnover → ↑ purines → hyperuricemia, gout risk.

18. → B) Phlebotomy

📌 First-line to lower Hct & blood viscosity.

19. → B) Spent phase

📌 Late PV → marrow fibrosis, cytopenias, splenomegaly.

20. → A) Transformation to acute leukemia

📌 Rare but most feared long-term complication of PV.

Done by Abdallah Alanzy