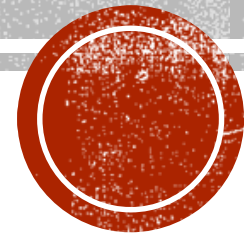


POLYCYTHEMIA

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DEFINITION

The difference between the two terms that when the number of the RBCs increased as a result of deficiency in hemoglobin, the mass will not increase in this case.

- Increase in total RBC mass above normal range
- Erythrocytosis: increased RBCs number

Fake one ▪ Relative polycythemia: secondary to decreased plasma volume (water deprivation, severe diarrhea, diuretics) **Not a disease related to the haematology**

- Primary: polycythemia vera (low erythropoietin, splenomegaly)

→ (ع تنوع كرات و تخریطوا حالتهم
منها)

-Polycythemia can be classified as the following:

- 1-Relative polycythemia.
- 2- Absolute polycythemia



ABSOLUTE POLYCYTHEMIA

- Absolute polycythemia: true increase in RBC mass, secondary to increased BM production
- Can be primary or secondary
- Primary: autonomous high bone marrow production (polycythemia vera), erythropoietin is low *Suppressed by negative feedback*
- Secondary: systemic hypoxia → high erythropoietin → increased erythropoiesis

Bone marrow

Hormon

Patients have another disease causes secondary type



SECONDARY POLYCYTHEMIA

Its causes :

- Adaptive: living in high altitude, cyanotic heart disease, chronic pulmonary diseases, sleep apnea
 Will result in life long hypoxia
 Example on chronic lung disease
- Paraneoplastic: renal cancer, liver cancer (as erythropoietin is secreted mainly by kidneys and a bit by liver)
- Surreptitious (blood doping): endurance athletes many athletes take EPO as a supplement
- Alcohol: frequent urination, depressed respiration
- Smoking Cause hypoxia and lung disease
- In secondary polycythemia: no splenomegaly
 as well as, the erythropoietin is high

Here if we treat the cause the patient will return to the normal state (reversible)



POLYCYTHEMIA VERA

Primary type

- Myeloproliferative neoplasm
- Mutation in tyrosine kinase JAK2 in bone marrow stem cells
 - This mutation presents in 99% of patients
- Normally acts in the signaling pathway of erythropoietin receptor and other growth factor receptors
 - While this mutation stimulates this pathway
 - This mutation presents in the earliest cell (stem cell) so all BM cells will carry it
- Hematopoietic cells become less dependent on growth factors
- Excessive proliferation of erythroid, myeloid cells and megakaryocytes (panmyelosis) Pan means all
- Erythrocytosis is most prominent, results in polycythemia
- Splenomegaly is common
 - Because it is a neoplastic disease, so the neoplastic cells go into the circulating blood and we see them in the spleen resulting in splenomegaly



SYMPTOMS OF POLYCYTHEMIA

Primary and secondary

Redness

- Plethora/ cyanosis **Bluish of the skin because of hypoxia**
- Headache and dizziness (from hypertension) **Because of the increase of blood mass**
- **Slow circulation and hyperviscosity cause cyanosis, blurred vision, tissue ischemia**
- **Thrombosis, or bleeding** (disturbed function of vWF)

Because the blood is full with erythrocytes, and they slower than the normal when they move resulting in viscous blood which affects the organs such as retina

Thrombosis
because of
increasing
platelets
and
bleeding
because of
abnormal
function of
platelets

In polycythemia vera: similar symptoms plus:

- **Pruritus** (aquagenic) **Because of increased WBCs and histamine**
- **Peptic ulcer** → Means it caused by water
- **Secondary gout** (arthritis, kidney stones, tophi)
- **Chronic disease** هذا وصف المرض
- **Spent phase:** occurs after an interval of **10 years of symptoms**, **BM becomes fibrotic**, **hematopoiesis shifts to spleen**
- **Blast crisis:** transformation to acute myeloid leukemia (rare)



LABORATORY FINDINGS OF POLYCYTHEMIA

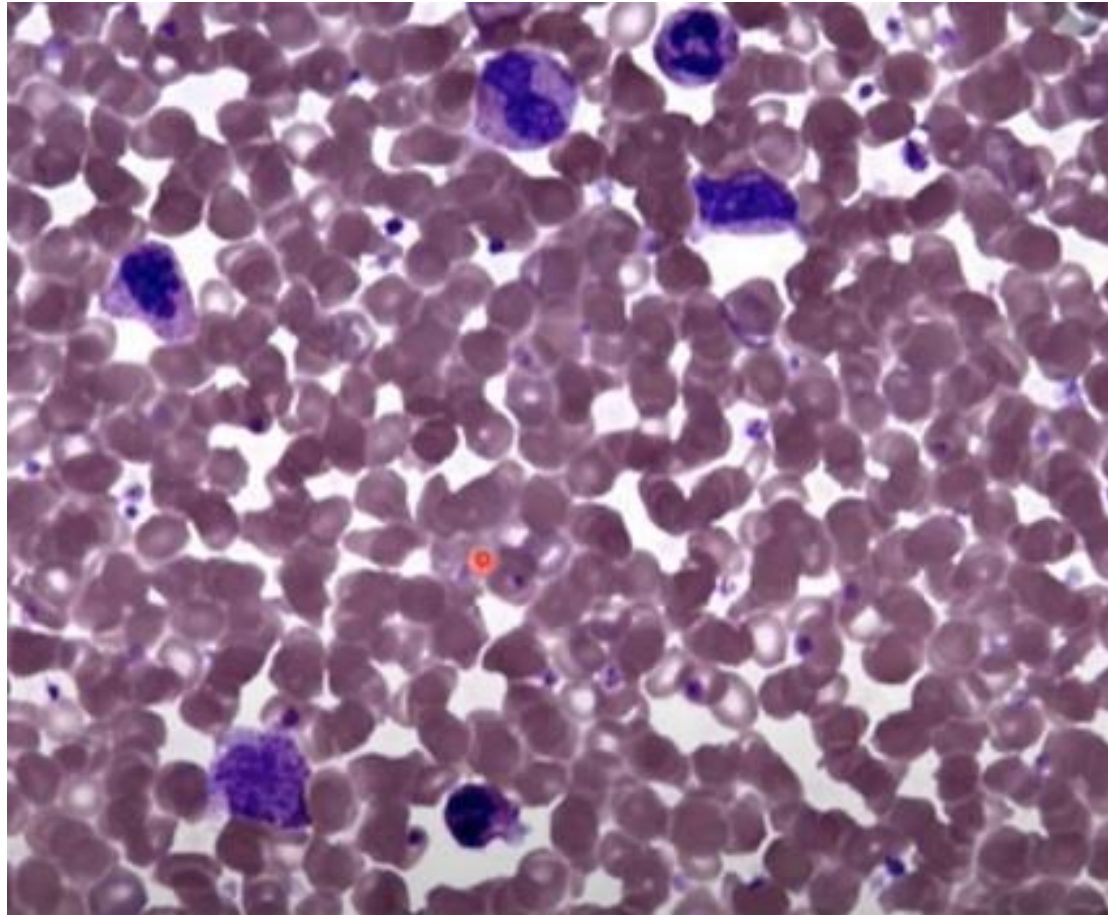
This is persistent

- High hemoglobin concentration (>16.5 g/dL in men, 16 in women) and high hematocrit (>49% in men, 48% in women)
- High RBCs count Erythrocytoses
- These tests might be masked if iron deficiency develops

In polycythemia vera: additional findings:

- Leukocytosis and thrombocytosis are common
- JAK2 mutation
- Low erythropoietin level
- Hypercellular bone marrow with panmyelosis Full with cells





- Peripheral blood smear in polycythemia: packed RBCs Very crowded

