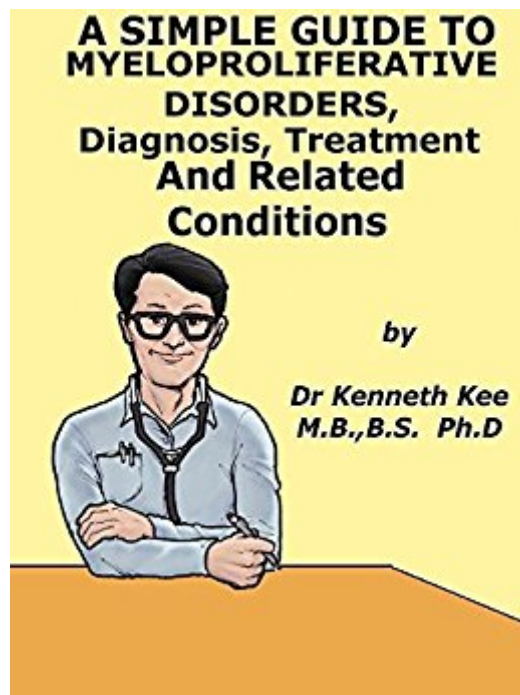


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# A Simple Guide To Myeloproliferative Disorders, Diagnosis, Treatment And Related Diseases (A Simple Guide To Medical Conditions)



## Synopsis

Chapter 1 Myeloproliferative Diseases

Myeloproliferative Diseases are rare in Singapore but Polycythemia vera and chronic leukemia are present in patients here. The treatment of Myeloproliferative Diseases is helpful in the knowledge of these diseases and the best way to treat them.

What are Myeloproliferative Diseases? Myeloproliferative Diseases are a group of hematological diseases characterized by varying degree of uncontrolled proliferation of 1 or more hematological cell lines in the peripheral blood, different from acute leukemia. The patient may undergo transition from 1 form of disease to another or go on to granulocytic leukemia. There are 4 diseases under this condition: 1. Chronic myelogenous leukemia (CML) 2. Polycythemia vera (PV); 3. Essential thrombocythemia (ET); 4. Myelofibrosis (MF), which is also known as agnogenic myeloid metaplasia (AMM). In 2002, the World Health Organization (WHO) had added 2 other diseases to this group of diseases: 5. Chronic neutrophilic leukemia (CNL) 6. Chronic eosinophilic leukemia (CEL)/Hypereosinophilic syndrome (HES).

What are the causes of Myeloproliferative Diseases? Causes: The cause is the proliferation of one of hematological condition underlying the disease.

1. Polycythemia rubra vera: There is the proliferation of all the bone marrow cells elements. There is increased number of red blood cells, increased hemocrit and increased hemoglobin.
2. Myelofibrosis (agnogenic myeloid metaplasia): There is the proliferation of fibroblasts in the bone marrow accompanied by extramedullary hematopoiesis.
3. Essential thrombocythemia: There is the increase of platelets.
4. Chronic myelogenous leukemia: Increased production of neutrophils, eosinophils, basophils, monocytes and thrombocytes occurs in the chronic phase of chronic myelogenous leukemia.
5. Chronic neutrophilic leukemia: There is the increase of neutrophils.
6. Chronic eosinophilic leukemia: There is the increase of eosinophils.

All are variants of a common disease process, possibly from the malignant transformation of a single stem cell. Fibrotic tissue (connective tissue) takes the place of the bone marrow and anemia may develop as the disease progresses. Extramedullary hematopoiesis occurs also in the liver and spleen causing hepatosplenomegaly.

What are the symptoms of Myeloproliferative diseases? The onset of symptoms are insidious occurring in older patients. Myelofibrosis is frequently diagnosed by the discovery of an enlarged spleen in asymptomatic patients. Common symptoms are:

1. Fatigue
2. Weight loss
3. Pallor (except in patients with polycythemia vera)
4. Ankle edema
5. Abdominal discomfort due to the presence of splenomegaly: This is most common in chronic myelogenous leukemia and agnogenic myeloid metaplasia.
6. Left upper quadrant and left shoulder pain as a consequence of splenic infarction and perisplenitis.

Other symptoms are:

1. Plethora secondary to polycythemia
2. Petechiae or ecchymosis
3. Palpable spleen and/or liver
4. Easy bruising, bleeding,

or symptoms of thrombosis<sup>5</sup>. Swollen, painful joint secondary to gouty arthritis that is due to hyperuricemia<sup>6</sup>. Priapism, tinnitus, or stupor from leukostasis<sup>7</sup>. Headache<sup>8</sup>. Dizziness and vertigo<sup>9</sup>. Visual disturbances<sup>10</sup>. General ill health

Investigations:

1. Full blood count (FBC), differential count with microscopic exam of the peripheral smear
2. Red blood cell mass study: True versus spurious polycythemia
3. Serum uric acid level is usually high in polycythemia
4. Bone marrow aspiration and biopsy with cytogenetic studies
5. Bone marrow histology shows the presence of hypercellularity in most of these disorders.

Bone marrow fibrosis is shown on the reticulin stain in myelofibrosis. Bone marrow fibrosis is also found in the later phase of chronic myelogenous leukemia and polycythemia vera.

Introduction

- 1 Myeloproliferative Diso

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