Myeloproliferative Disorders

Brian Meier, MD, MA[a], John H. Burton, MD[b,*]

KEYWORDS

• Myeloproliferative disorders • Essential thrombocythemia • Polycythemia vera
• Chronic myelogenous leukemia • Primary myelofibrosis

KEY POINTS

- The emergency provider (EP) generally encounters myeloproliferative disorders (MPNs) in 1 of 2 ways: as striking laboratory abnormalities of seeming unknown consequence, or in previously diagnosed patients presenting with complications.
- Rapid hydration, transfusion, cytoreduction, and early hematology consultation can be lifesaving.
- It is not uncommon for an MPN to initially be considered by the EP after notification from the hospital laboratory that an emergency department patient has an elevated cell count on a complete blood count assay.

INTRODUCTION: NATURE OF THE PROBLEM

It is not uncommon for a myeloproliferative disorder (MPN) to initially be considered by the emergency provider (EP) after notification from the hospital laboratory that an emergency department (ED) patient has an elevated cell count on a complete blood count assay. This finding may take the form of an elevation of a single cell line (eg, red cells, white cells, or platelets). Alternatively, all or multiple cell lines may be elevated in the patient's laboratory values.

When a patient with an elevated cell count presents to the EP, an MPN may often enter the differential diagnosis. Particular consideration should be given to the most common myeloproliferative neoplasms: essential thrombocythemia (ET), polycythemia vera (PV), chronic myelogenous leukemia (CML), and primary myelofibrosis (PMF).[1] Myeloproliferative neoplasms are characterized by normal bone marrow with subsequently terminal myeloid expansion in the peripheral blood, leading to pathologically increased numbers of one or more cell lines.[2]

This article originally appeared in Emergency Medicine Clinics of North America, Volume 32, Issue 3, August 2014.
[a] Department of Emergency Medicine, Carilion Clinic, 525 Janette Avenue Southwest, Roanoke, VA 24016, USA; [b] Department of Emergency Medicine, Carilion Clinic, PO Box 13367, Roanoke, VA 24033, USA
* Corresponding author.
E-mail address: JHBurton@carilionclinic.org

Hematol Oncol Clin N Am 31 (2017) 1029–1044
http://dx.doi.org/10.1016/j.hoc.2017.08.007 hemonc.theclinics.com
0889-8588/17/© 2017 Elsevier Inc. All rights reserved.

The entities that are classified as MPNs were first described by Vasquez in 1892. He noted a patient with erythrocytosis and splenomegaly, whom he rightly suggested as suffering from a hemoproliferative mechanism.[3] The entities now known as essential thrombocytopenia and primary myelofibrosis have been described as separate clinical entities. In 1951, Dameshek described these seemingly separate disorders as interrelated and proffered the concept of myeloproliferative syndromes.[4]

Work over the ensuing 50 years led to great advances in the understanding of the factors that influence hemoproliferation. This culminated in the work by Levine and colleagues[3] in 2005, which identified a tyrosine kinase mutation in the common JAK-2 allele. This rendered the best explanation to date as to how these disorders ensue. Since that time, a multitude of other cytogenetic abnormalities have been investigated, none of which have proved to be definitive.

DEFINITIONS

In 2008, the World Health Organization altered the classification system of myeloid neoplasms. These entities are now divided into the categories listed in **Box 1**.

The major determination is made between those entities that are considered myelodysplastic from those that are considered myeloproliferative. Myelodysplasia is defined by dysplastic, or abnormal, bone marrow resulting in cytopenia of varying degrees due to intramedullary apoptosis.[5] MPNs, in contrast, are notable for normal bone marrow findings with increased cell line count(s) in the peripheral blood.

Box 1
World Health Organization (WHO) classification of myeloproliferative neoplasms

- Myeloproliferative neoplasms (MPN)
  - Chronic myelogenous leukemia (CML)
  - Polycythemia vera (PV)
  - Essential thrombocythemia, also known as essential thrombocytosis (ET)
  - Primary myelofibrosis (PMF)
  - Chronic neutrophilic leukemia (CNL)
  - Chronic eosinophilic leukemia, not otherwise specified (CEL-NOS)
  - Mast cell disease (MCD)
  - MPN, unclassifiable
- Myelodysplastic syndromes (MDS)
- Myelodysplastic syndrome/myeloproliferative neoplasm (MDS/MPN)
  - Chronic myelomonocytic leukemia (CMML)
  - Juvenile myelomonocytic leukemia (JMML)
  - Atypical chronic myeloid leukemia, BCR-ABL–negative (aCML)
  - MDS/MPN, unclassifiable
- Acute myeloid leukemia (AML)
- Myeloid and lymphoid neoplasms with eosinophilia and abnormalities of PDGFRA, PDGFRB, and FGFR1

Data from Tefferi A, Thiele J, Vardiman JW. The 2008 World Health Organization classification system for myeloproliferative neoplasms. Cancer 2009;115(17):3842–7.

Download English Version:

https://daneshyari.com/en/article/8734010

Download Persian Version:

https://daneshyari.com/article/8734010

Daneshyari.com