

Hematology for Practitioners

Edited by
Marshall A. Lichtman, M.D.

HEMATOLOGY FOR PRACTITIONERS

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Hematology for Practitioners

Robert Inslee Weed

1928–1976

This book is dedicated to the memory of Robert I. Weed, Professor of Medicine and of Radiation Biology and Biophysics and Chief of the Hematology Unit at the University of Rochester Medical Center and the School of Medicine and Dentistry. During his too short professional life his brilliance and dedication to learning kept tilled the soil in which new knowledge could blossom. As a scientist, educator, and physician, his imaginative ideas, stimulating discourse, and clinical skills inspired his students and peers.

Preface

The physician has access to several excellent textbooks of hematology. These texts describe in a comprehensive fashion the anatomy, physiology, biochemistry, and pathology of blood-forming organs, blood cells, and blood proteins. They are excellent resources for students, basic scientists, and physicians, particularly specialists. Although valuable for the student and specialist, the comprehensive subspecialty textbook is difficult for the practitioner to use prospectively as he cares for patients. Introductory texts, on the other hand, often emphasize physiologic and biochemical aspects of the blood and are oriented, therefore, toward the student.

The general physician does not share the luxuries of the medical student who has neither the responsibility for decisions nor the pressures of time limitations in which to make them; of the basic scientist who does not have to make decisions that affect the life of a patient; or of the specialist who is versed in the measurements he uses often, in the uncommon diseases he sees frequently, and in the difficult therapeutic decisions that he faces repeatedly.

General physicians may find hematologic problems difficult because their diagnosis is dependent on laboratory signs rather than physical signs. Moreover, the laboratory tests, such as the blood or marrow smear used to validate the presence of many hematologic diseases, may not be examined frequently by the physician, who thus may not have developed high competency with their interpretation.

In this text we have tried to maximize the information that is relevant for the diagnosis of major hematologic diseases. We have done this from the vantage point of the key or most consistent sign that a blood disease produces. In this way we hope to facilitate the use of the book by the general physician, who can enter the book at the chapter or chapters that discuss the major hematologic sign or signs manifested by the patient. This also imposes on the physician the requirement to crystallize the chief sign. For example, he should decide whether an isolated cytopenia or multi-cytopenia is present. If anemia alone is present, the pathophysiology of the anemia—reticulocytopenic, implying marrow failure, or reticulocytotic, implying hemolysis or hemorrhage—needs to be determined.

Treatment of patients with hematologic diseases can also be troublesome for the general physician. The management of hematology patients who require drugs with a high toxic-to-therapeutic ratio, component transfusion therapy, repeated evaluation of marrow examinations, and intensive care is difficult since the infrequency of these diseases prevents experience and thus limits wisdom in making decisions. Treatment is often orchestrated by the specialist. However, the general physician can play a valuable role in the care of the patient. Too often this role is abdicated because of an unfamiliarity with the essentials of the natural history of the disease, the principles of treatment, and the effects of drugs that are used infrequently.

For these reasons we have included chapters on surgical, medical, and radiotherapeutic treatments of hematologic disorders. Knowledge of the principles, utility, and consequences of therapy will help the general physician participate in the care of a patient who may have to travel long distances to be evaluated by a specialist.

The Hematology Unit at The University of Rochester School of Medicine and Dentistry and some of its alumni delivered a course in hematology sponsored by the American College of Physicians in June 1976. Following this course, we proposed to prepare a book in which the chapters were entitled by common signs of hematologic diseases. In general, each sign of a hematologic disease may be pursued by a parsimonious diagnostic strategy. Our treatment of these disorders is meant neither to underestimate the complexities of human biology and the difficulties of diagnosis and management nor to foster medicine by algorithm. Rather we have tried to use the physiologic, biochemical, and biophysical knowledge that has accrued in the study of hematologic diseases to instruct in the processes of diagnosis. There are many texts and articles in which additional facts about hematologic diseases can be found. We have tried to provide a practical framework for the diagnosis and management of hematologic disorders for the unspecialized physician and the nonhematologist.

M. A. L.

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Mrs. Carol B. Weed and Stase Mickys provided invaluable help in the preparation of the manuscripts for publication. Jean A. Shafer provided the micrographs of blood and marrow.

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NOTICE

The indications and dosages of all drugs in this book have been recommended in the medical literature and conform to practices of the general medical community at The University of Rochester School of Medicine and Dentistry. The medications described do not necessarily have specific approval by the Food and Drug Administration for use in the diseases and dosages for which they are recommended. The package insert for each drug should be consulted for use and dosage as approved by the FDA. Because standards for usage change, it is advisable to keep abreast of revised recommendations, particularly those concerning new drugs.

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I An Approach to the Diagnosis of Reduced Blood Cell Counts

Marshall A. Lichtman

One of the practical results of the increase in our understanding of organ pathology is the ability to arrive at a conclusion regarding the pathogenesis of disease. Often, that ability allows a physician to narrow his diagnostic considerations and to select laboratory tests in a more precise manner. If one could promptly decide whether a patient was suffering from an infectious, inflammatory, neoplastic, toxic, or degenerative disease of an organ, using simple techniques, further diagnostic efforts could be focused and thus made more informative and cost efficient.

The history and physical examination are important in the approach to cytopenias. A history of excessive menses, a folate poor diet, alcohol abuse, drug ingestion, a family history of anemia, and family roots in geographical areas where red cell enzyme or hemoglobin abnormalities are prevalent represent a few key findings which may provide clues to the etiology of the disturbance.

Usually, clinical laboratory tests are required to achieve a firm diagnosis. Simple tests are useful in arriving at a diagnosis. In hematopoietic disorders, especially cytopenias, the pathogenesis of the disturbance frequently can be discovered rapidly with studies that can be performed in a physician's office. The ability to decide that a cytopenia is a result of either decreased production of cells, increased destruction of cells, or loss of cells allows the physician to select the tests most likely to identify the causative factor in the cytopenia.

IDENTIFICATION OF AN ISOLATED CYTOPENIA

Automated techniques provide a measurement of the hemoglobin concentration, red cell count, packed cell volume (PCV), and leukocyte count. The blood smear provides the differential count of leukocytes and an estimate of platelet numbers. Those studies should be completed in order to define whether an isolated anemia, granulocytopenia, or thrombocytopenia is present. Bicytopenia and tricytopenia have different differential diagnoses and may require different diagnostic studies.

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