COLLAGEN DISEASES

Including

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DERMATOMYOSITIS • SYSTEMIC SCLERODERMA
THROMBOTIC THROMBOCYTOPENIC PURPURA

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TO

Arlie V. Bock

D. Bruce Dill

and the late

Lawrence J. Henderson

Physician, Physiologist and Philosopher, respectively. In spite of a great difference in personality, each one was to me an outstanding individual, a fine teacher and an ever-loyal friend.

JHT

Preface

However other writers of monographs in medicine may proceed, in my own case the Preface, always prepared last, marks a pleasant cessation of labors. With more satisfaction than probably is warranted, I turn to the compilation of an index and eventually to proofreading, the culmination of twenty years of sporadic awareness of the need for a concise monograph on the disorders described herein, and the final result of eighteen months of reference work, writing and rewriting.

Twenty years of sporadic awareness refers to a brief exchange of correspondence with the late Dr. Campbell P. Howard, one-time pupil of William Osler in Baltimore, when he was Professor of Medicine in McGill University at the peak of his professional career. It was my privilege to feel the warmth of the personality of "C.P.H." only through his written words, but I can vouch for his eminence as a teacher and clinician while entrusted with the Department of Medicine at the University of Iowa, only a few miles by dirt road from my birthplace. Although the correspondence began in 1936, the origin of a common interest may be traced to the preceding year, when, at the age of sixty, Dr. Howard, upon recognizing for the first time a case of scleroderma with calcinosis, described his feelings as follows: "To a student of medicine a new clinical experience always gives a thrill. Even the most blasé hospital physician is learning something new each day though he may not be prepared to admit it; certainly at least once a month he is faced by some problem that has not confronted him before. Such was my position early in January, when on making my daily ward visit, I was

shown this patient whose case report I am about to relate; I realized that the condition was a new one to me."

The quotation above is an excerpt from the introductory paragraph to a paper presented in 1935 before the Association of American Physicians* entitled, "A Case of Early Scleroderma with Calcinosis." It should be noted also that before the same scientific assembly Dr. George Baehr presented a communication entitled, "A Diffuse Disease of the Peripheral Circulation (Usually Associated with Lupus Erythematosus and Endocarditis)." No discussion followed Dr. Howard's presentation, but several queries were addressed to Dr. Baehr. Dr. Joseph H. Pratt of Boston, inquired: "I wish Dr. Baehr would tell us if he is able to make the diagnosis of the disease [lupus] in the absence of skin lesions. If so, I am especially interested to know how he would distinguish it from rheumatoid arthritis." Several comments were made by Dr. Emanuel Libman of New York, who displayed a great interest in this communication.

Thus far in the Preface, scleroderma and lupus erythematosus have been introduced as well as the difficulty encountered at times in distinguishing lupus from rheumatoid arthritis. Dr. Howard used the term, "scleroderma with calcinosis" to describe his case, but he might have committed an error similar to mine, in describing a case of "dermatomyositis with scleroderma and calcinosis . . . ,"[*372*] because the differentiation between dermatomyositis and scleroderma, in patients with calcinosis, has not been defined clearly. Yet one other "collagen disorder" had attracted the interest of Dr. Howard a decade earlier. He reviewed the literature on periarteritis nodosa in 1925 [*216*] at the same time that Moschowitz [*579*] described a case of thrombotic thrombocytopenic purpura.

At the time of his death in 1936 Dr. Howard had prepared a fine bibliography on the subject of calcinosis, which supplemented in some instances and duplicated in others, a similar project that I had undertaken. Only a modicum of

* Transactions of the Association of American Physician, vol. 50, 1935.

progress could be documented before World War II brought an effective halt to my efforts. Following the war, the increasing incidence of several "rare" collagen disorders was directly responsible for the cessation of daydreaming about the preparation of the manuscript and for the stimulus to start working upon it. The results constitute the chapters that follow on Systemic Lupus Erythematosus, Polyarteritis, Dermatomyositis, Systemic Scleroderma and Thrombotic Thrombocytopenic Purpura.

It is always a pleasure to acknowledge assistance from persons and institutions who have lent a helping hand. Many times also the recipients of this acknowledgment are pleased to be so recognized. I have even assumed at times that institutions, such as hospitals and medical schools, cold impersonal structures on the outside, may derive some inward sense of appreciation when they are the object of acknowledgment.

Dr. Ricardo Ferrandis, a Castilian, studying in America and currently a Training Fellow in Arthritis, helped tremendously in the library work. Somewhat more than five times as many articles were read as are listed in the bibliography. In most instances decision regarding acceptance or rejection was made after each of us had scanned the original one or more times. But Dr. Ferrandis' work on the manuscript is not over. He has already begun to translate it into Spanish for the Latin American countries. I believe that as North Americans we are charged with the responsibility of helping, whenever we are able, to exchange suitable medical information with our associates South of the Border. I hope this, the latest of several efforts on my part to do so, will not be the last.

The pathologic material was made available by my colleagues, Drs. Kornel Terplan and Dorothy Shaver in the Department of Pathology, and Drs. Stuart Vaughan, Raymond Kibler and Miss Jeanette Corbett in the Department of Clinical Pathology. The excellence of the roentgenography is due to the interest of Dr. Gordon Culver of the Department

of Roentgenology. Selected material for reproduction has been provided by Dr. Earl Osborne of the Department of Dermatology, by Dr. Joseph Acquilina of the Buffalo Veterans Administration Hospital, by Dr. Ashton Welsh of Cincinnati, and by Dr. Salvatore Latona of Niagara Falls.

The following physicians have provided clinical material either from their private practice or from their ward service in the hospital: Drs. Abraham H. Aaron, Carl E. Arbesman, Grovenor W. Bissell, Marvin L. Bloom, Jay I. Evans, Stephen A. Graczyk, Raymond S. Kibler, George F. Koepf, William F. Lipp, Morton H. Lipsitz, L. Maxwell Lockie, Elmer T. McGroder, Harry M. Murphy, M. Luther Musselman, Bernard M. Norcross, Francis W. O'Donnell, Howard Osgood, Harold M. Robins, Nelson G. Russell, Jr., Nelson G. Russell, Sr., Frederick T. Schnatz, Max A. Schneider, Stuart L. Vaughan, Samuel A. Vogel, Joseph E. Anderson of Olean, Alfred E. Dooley of Auburn, Walter S. Finken of Bradford, and John E. Thompson of Youngsville, Pennsylvania. Except where noted, the above-mentioned are members of the medical community of Buffalo. This is a long list, but the appreciation is conveyed individually and sincerely. I am indebted also to my secretaries, Mrs. Geri Kassirer and Miss Barbara Allan for painstaking copying, and to Mr. Raymond M. Verrill, of the English Department of Nichols School, for typereading.

I am especially grateful to Sharp & Dohme, Division of Merck & Co., for permission to reproduce the color figures and several of the black and white illustrations. The color plates (pages 1–8) appeared originally in three issues of *Seminar* in order to illustrate articles on similar subjects that were prepared prior to this manuscript. Color plates, particularly appropriate for medical reports, are prohibitive in cost. Lacking special funds, the arrangement for reproducing them would have been impossible without the aforementioned support.

Acknowledgment to institutions, as noted above, referred to essential contributions that each has made by virtue

of the stature enjoyed in their respective communities. A number of patients with dermatomyositis and scleroderma were followed at the Massachusetts General Hospital in the 1930's. Since 1946 the clinical material has been drawn largely from the Buffalo General Hospital. A few cases have been seen at the Veterans Administration Hospital in Buffalo, and isolated cases in other hospitals in Western New York State. Finally, the School of Medicine at the University of Buffalo has been the academic mine of precious metals from which physicians and institutions pursue their respective missions in these hills of learning, occupying varying degrees of independence, but never losing touch with the mother lode.

This monograph has been prepared without any specific grants-in-aid. However, several persons and Foundations have contributed directly to the current research program in the Department of Medicine, from which this monograph comes. I am pleased, therefore, to acknowledge financial support from the following: Mrs. Kathleen B. S. Chard, Cazenovia, New York; Mr. Alton Wood, Eden, New York; The National Institutes of Health, Bethesda, Maryland; The John A. Hartford Foundation, New York City; The Western New York Chapter of the Arthritis and Rheumatism Foundation; and the Masonic Foundation for Medical Research and Human Welfare.

JHT

Eden, New York
July, 1955

COLOR PLATES

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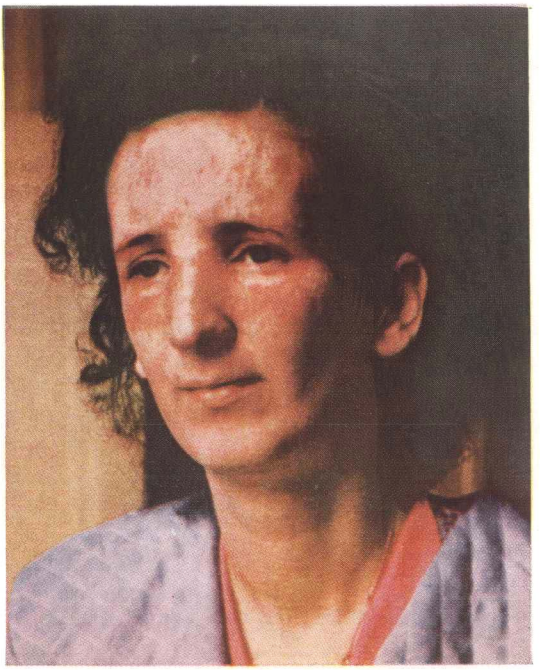
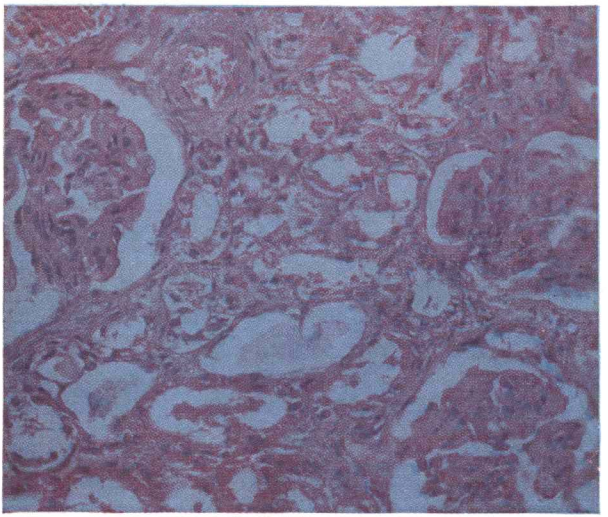


PLATE I.—Butterfly rash on the face of a patient with SLE.



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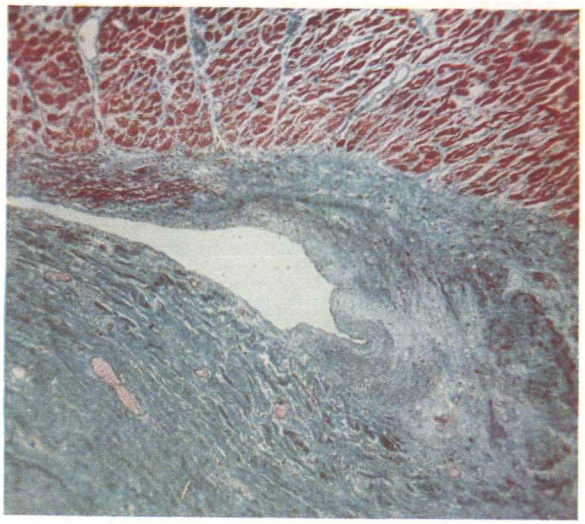

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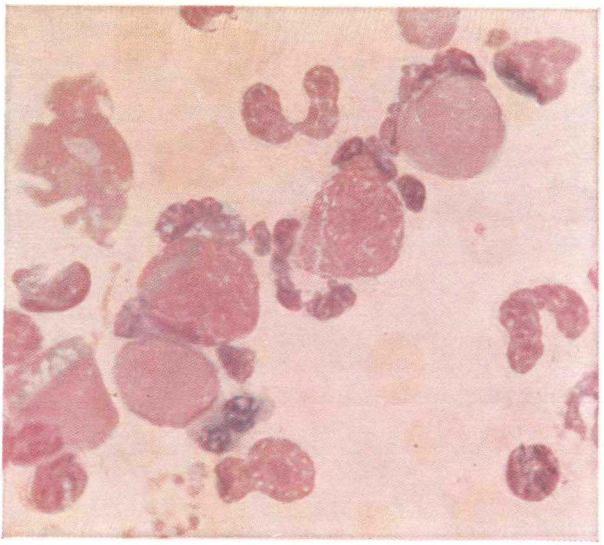

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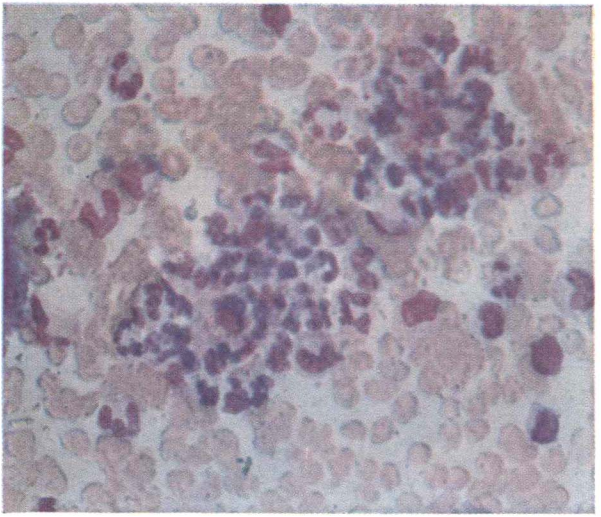

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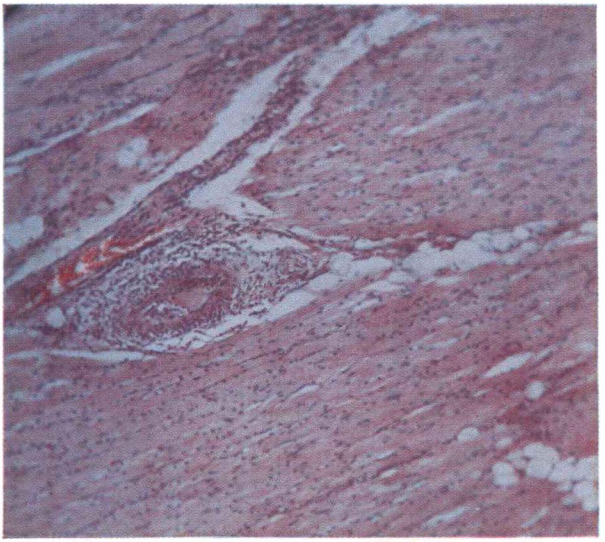
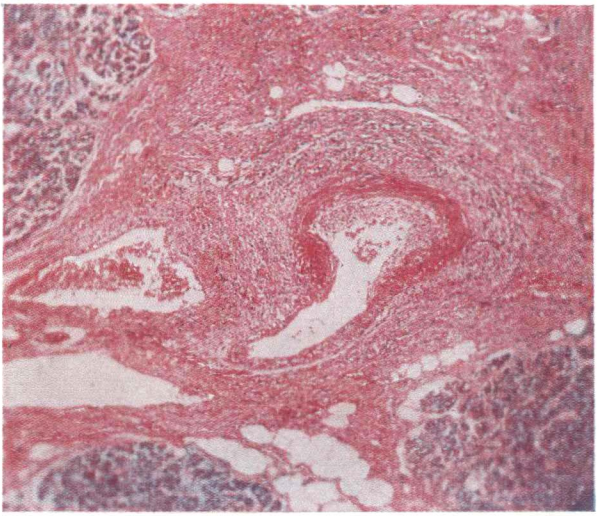


PLATE VI.—Section of a muscle removed at biopsy from a patient with polyarteritis. There is acute fibrinoid necrosis of the media in an arteriole surrounded by dense cellular infiltration. The inflammation extends between the adjoining muscle bundles. × 96.



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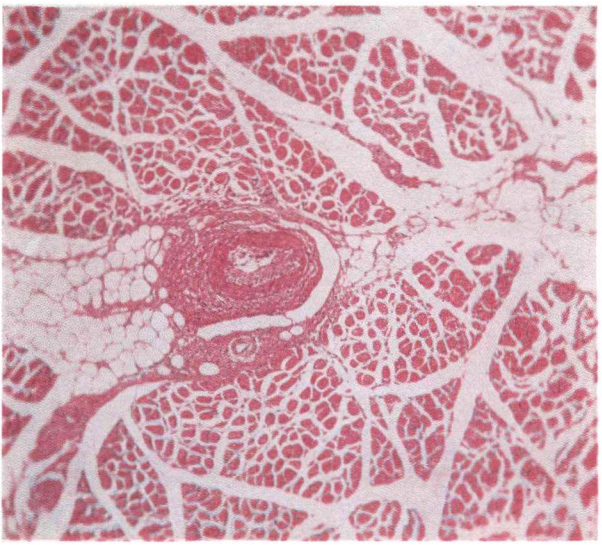

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