Systemic INFLAMMATORY DISEASE AND THE EYE

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WRIGHT

Systemic Inflammatory Disease and the Eye

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WRIGHT
Bristol
1987

Published under the Wright imprint by
IOP Publishing Limited.
Techno House, Redcliffe Way, Bristol BS1 6NX

British Library Cataloguing in Publication Data
Dinning, W.
Systemic inflammatory disease and the eye
1. Ocular manifestations of general diseases
I. Title
616.07'2 RE65

ISBN 0 7236 0777 X

Typeset by
Bath Typesetting Limited

Printed in Great Britain by
The Bath Press, Lower Bristol Road, Bath BA2 3BL

SYSTEMIC INFLAMMATORY DISEASE AND THE EYE

Dedication to my mother and father

Preface

Several years ago I began a programme of reading with two objectives in mind. The first was to achieve an overview of the many systemic conditions associated with ocular inflammation. The second and more alluring one was to gain substantial insights into the reasons for ocular involvement in some conditions and not in others.

Encouraged by colleagues and convinced that others would find it both helpful and interesting, I have here recorded the outcome of this pursuit. The book is aimed at the middle ground between internists and ophthalmologists. I hope it will provide ophthalmologists with an informed background to ocular inflammation, and also help internists gain a better understanding of the nature of the ocular lesions. The latter may find the introductory chapter on signs of ocular inflammation of particular value.

While the pursuit of my first objective has been personally very rewarding, I find myself frustrated in the second. I firmly believe that the necessary revelations will eventually be found in the study of immunopathology, but the torrent of information which is adding daily to our knowledge of the mechanisms of disease seems to generate scarcely a trickle about the true *cause* of disease. My present feelings are well expressed by Robert Musil:

> In these hundred years we have got to know ourselves and Nature and everything very much better, but the result is, so to speak, that whatever one gains in the way of order in matters of detail one loses again where the totality is concerned, so that what we get is more and more systems of order and less and less of order itself.
>
> *Der Mann ohne Eigenschaften*, 1930

My gratitude is due to the many colleagues who have been so generous with encouragement and constructive criticism, and to the medical librarians who, with their unfailing cheerfulness, patience and understanding always make me feel welcome in their domains.

W. J. D.

Acknowledgements

I am most grateful to the many colleagues who have provided illustrations for this book. Their names have been added to the appropriate captions.

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Foreword

by **G. Richard O'Connor** MD
Professor of Ophthalmology, Emeritus,
University of California, San Francisco

There has long been a need for a clinical text that would aid both internists and ophthalmologists in their struggle to diagnose and treat inflammatory disorders of the eye with efficiency and safety. William Dinning's latest literary effort, *Systemic Inflammatory Disease and the Eye*, provides just such a book.

In the past, ophthalmologists and internists have dealt with the subject patients as if the latter belonged to a discrete domain. For his part, the ophthalmologist has generally regarded the ocular inflammation as an isolated lesion that should be treated with corticosteroids. The possibility of coexistent, related systemic disease is often disregarded, particularly if the patient in question has no systemic complaint, or at best, a few minor aches and pains. It is now known that such patients may well have incipient rheumatoid disease that can be confirmed at an early stage only by laboratory tests or roentgenographic examinations. On the other side of the coin, immunologically compromised hosts, such as patients with early sarcoidosis or 'acquired immune deficiency syndrome' (AIDS), may have no systemic symptoms at all at the beginning of their disease. An ocular lesion may be the only manifestation of a potentially lethal systemic disease. To treat such a patient with orally administered corticosteroids without a thorough investigation might be extremely dangerous, particularly if the ocular lesion were attributable to an opportunistic pathogen such as *Candida albicans* or *Toxoplasma gondii*.

On his part, the internist, upon receiving a referred patient from a concerned ophthalmologist, will often subject the patient to a broad battery of laboratory tests, many of which have no relevance to the case at hand. Such mistakes, though generally harmless, are nevertheless the causes of needless expense and inconvenience. The internist often regards uveitis, for example, as a single disease. For lack of specific orientation from the referring ophthalmologist, he proceeds to order useless laboratory tests such as the ones for 'rheumatoid factor' in cases of focal chorioretinitis. The lesion in such cases is much more likely to be caused by an infectious organism, e.g., *Toxoplasma gondii* or *Treponema pallidum*. The internist may also order multiple tests for diabetes, hepatitis, and chronic renal disease, most of which net no useful information.

The major problem in both situations is a lack of familiarity with the applicable syndromes. Although one generally learns in medical school

the components of the various syndromes that relate eye diseases to systemic disorders, it is easy to forget some of these items. Recently acquired information, such as that dealing with AIDS, was never available to those of us who attended medical school several decades ago. In this book Dinning defines all the important syndromes relating to ocular inflammation. He describes their characteristic presentation, detailing the physical appearance and location of the lesions as well as their usual clinical course. Useful definitions of the terms frequently used by ophthalmologists to describe the lesions are presented in the first chapter. Dinning presents current and appropriate ideas about the common pathogenesis of the ocular and systemic lesions, although much of this material must still be regarded as theoretical. Lastly he provides cogent ideas about the treatment of patients suffering from the specific diseases under discussion.

Readers of this book will find the material accurately portrayed. The references are up-to-date and appropriately placed. The style of writing is that of an experienced medical observer and conscientious scholar.

Contents

Chapter one

The Signs of Ocular Inflammation

Physicians reading this book will meet many terms from the jargon of ophthalmology with which they may be unfamiliar. This chapter aims to explain and illustrate many words used to describe inflammatory signs in the eye. In addition, reference is made to pathological findings which may be relevant to the genesis of these lesions and their relationship to associated systemic disease.

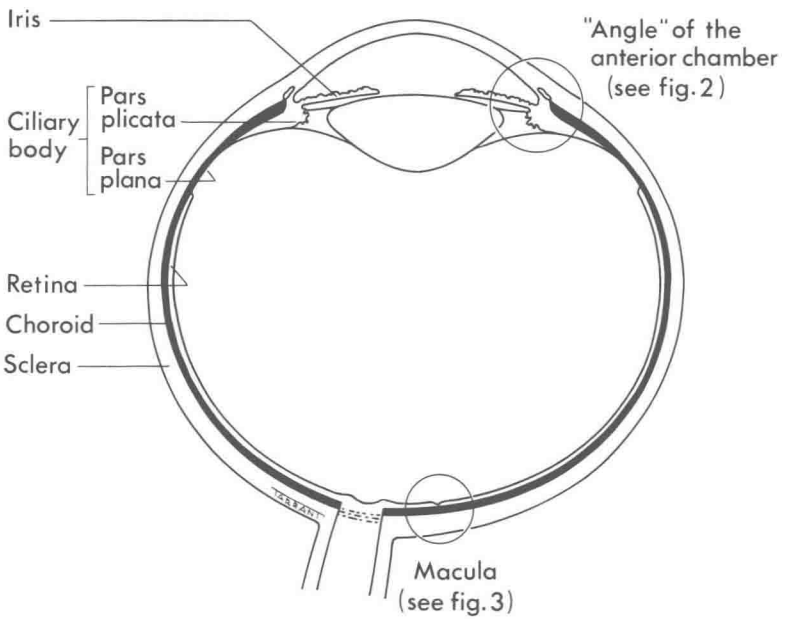

Figure 1 Horizontal section of the eye.

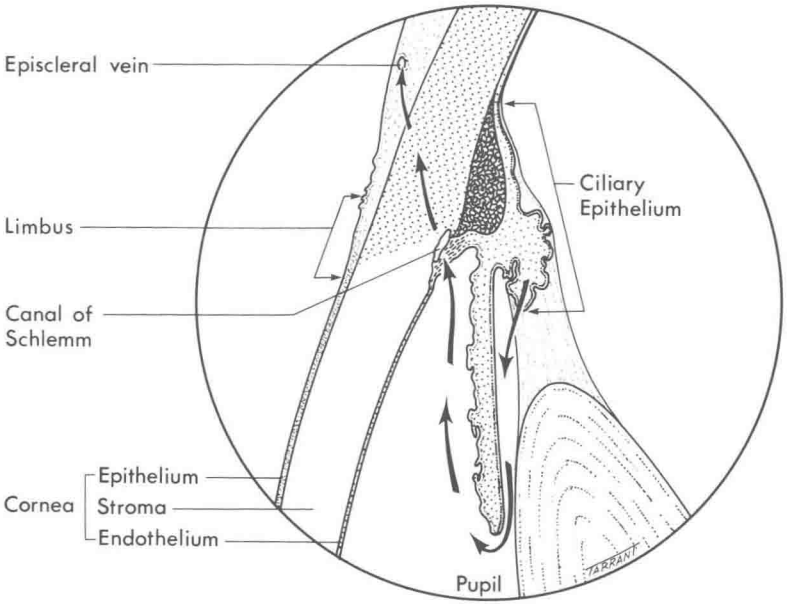

Figure 2 The angle of the anterior chamber showing the route of aqueous flow.

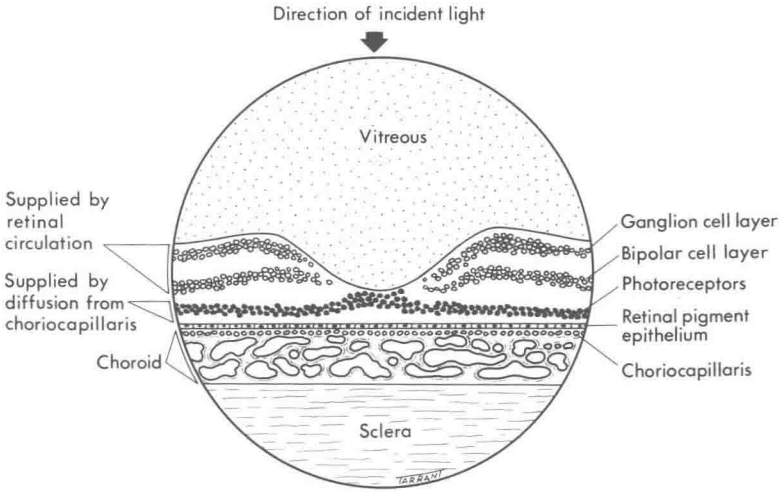

Figure 3 The retina at the macula.

Eyelids

Patients with scleroderma may have cicatricial contraction of the eyelids leading to poor closure and secondary exposure keratitis (q.v.).

Conjunctiva

(1) Conjunctivitis
(2) Nodules

1 Conjunctivitis

In *conjunctivitis* the redness extends uniformly from the edge of the cornea to the periphery of the eye. Only a segment of the eye may be affected. The dilatation of the superficial blood vessels is not normally confined to the conjunctiva of the eyeball but involves that of the lids as well. Unless there is secondary infection, any associated discharge is clear and watery.

2 Nodules

These occur in the conjunctiva in sarcoidosis. They are distinguished from common degenerative and cystic changes by their yellowish, solid, fleshy and somewhat vascular appearance. They occur most commonly in the lower fornix and on the plica semilunaris. They vary from one to several millimetres across, and may be single, multiple or confluent. (*Fig.* 4.)

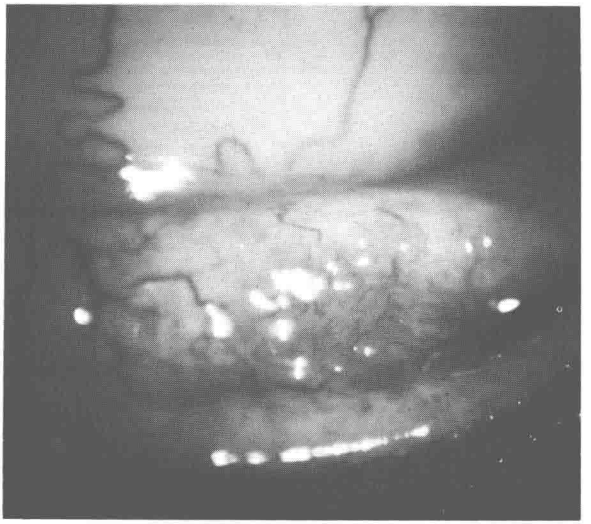

Figure 4 Sarcoid nodules in conjunctiva.
(Dr Robert Nussenblatt.)

Lacrimal Glands

In Sjögren's syndrome the lacrimal glands tend to be enlarged initially, becoming small and firm later in the disease. Enlarged glands may present a visible bulge under the outer part of the upper eyelid. The palpebral lobe of the gland can normally be brought into view by forceful elevation of the outer part of the eyelid while the patient looks down and inwards.

Episclera

Dilated episcleral vessels can be identified because the conjunctiva can be moved over them while they remain relatively fixed. Episcleritis usually involves only a segment of the surface of the eye, the redness contrasting with the adjacent white of the sclera. It is a deeper red than conjunctivitis.

Sclera

Scleritis is discussed fully in the chapter on rheumatoid arthritis. It may be diffuse or nodular. The conjunctiva is elevated over it, and the redness has a bluish tinge. Nodules tend to become yellow after the initial inflammation has begun to settle. (*Fig.* 5.)

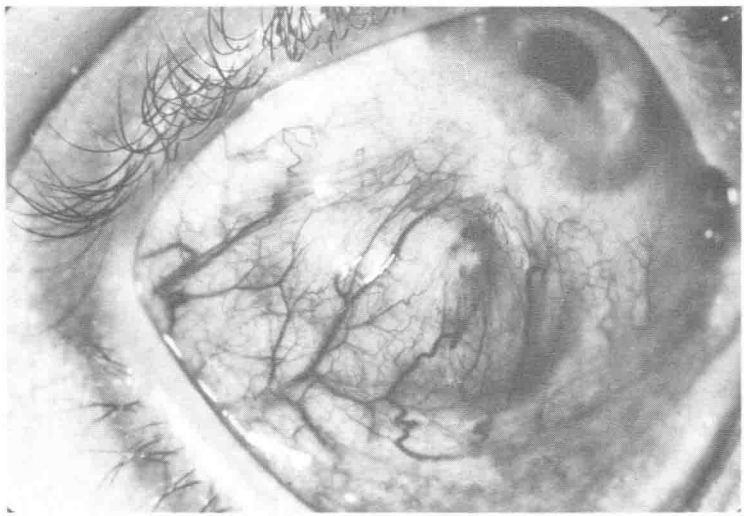

Figure 5 Sclerokeratitis.
(Dr Robert Nussenblatt.)

Ciliary Injection

Ciliary injection derives its name from the fact that the dilated vessels giving the classic circumcorneal blush are twigs from the anterior ciliary arteries, which anastomose within the major arterial circle of the iris, and ciliary efferent veins from the ciliary venous plexus. This circumcorneal redness is the hallmark of inflammation of the iris and ciliary body, but surface redness is confined to this area only in mild cases. It is often much more widespread, involving the episcleral and conjunctival vessels as well. In acute anterior uveitis the whole eye is often red. Sometimes the lids and even the side of the face are swollen and injected. (*Fig.* 6.)

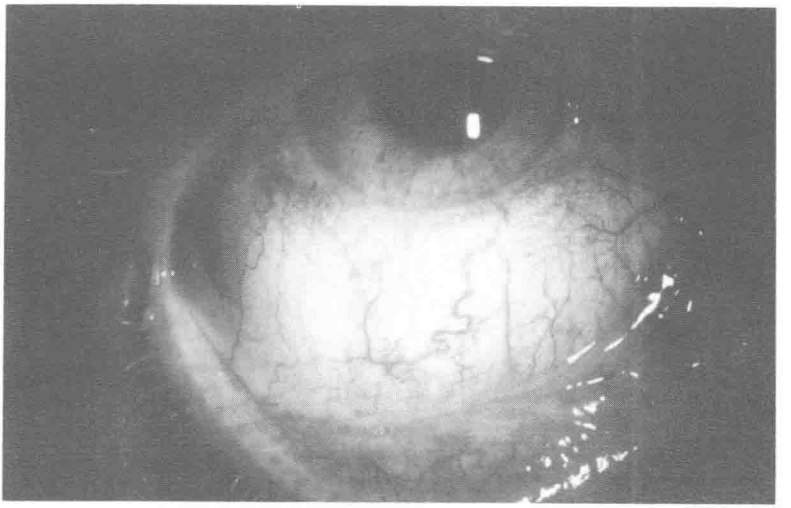

Figure 6 Mild ciliary injection.

Cornea

(1) Band keratopathy
(2) Keratoconjunctivitis sicca
(3) Keratitis
(4) Corneal endothelium
(5) Keratic precipitates

1 Band Keratopathy

In grossly deranged and shrunken eyes deposition of calcium salts beneath the corneal epithelium is common. It is not often seen in

sarcoidosis, but in the field of ocular inflammatory disease is most commonly met with in the uveitis associated with juvenile chronic polyarthritis.

A frosted glass appearance is first detectable near the limbus of the cornea at 9 and 3 o'clock, but separated from it by a clear zone. These areas become denser and extend horizontally towards each other. There may be circular clear areas within the zones of opacity. They may join across the visual axis and produce visual impairment. Symptoms are usually the result of small ulcers produced when calcareous fragments flake away. These areas become rapidly reepithelialized.

Pathological examination shows granular deposits of calcium along Bowman's membrane and a layer of fibrous tissue between Bowman's membrane and the corneal epithelium.

In cases of chronic uveitis that develop band keratopathy there is presumably a defect of corneal nutrition. The interpalpebral location suggests a role for temperature, hydration and gas exchange. (*Figs.* 7 and 8.)

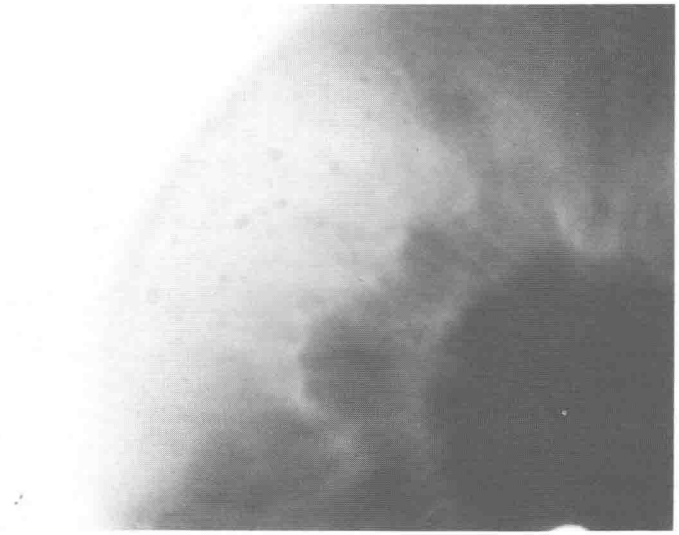

Figure 7 Mild band keratopathy.

2 *Keratoconjunctivitis Sicca*

When the eye is dry the cornea loses its lustre. Mucus tends to precipitate from the tears and forms strands in the lower conjunctival fornix. It may adhere in blobs to the epithelium and form filaments with tags of epithelium.

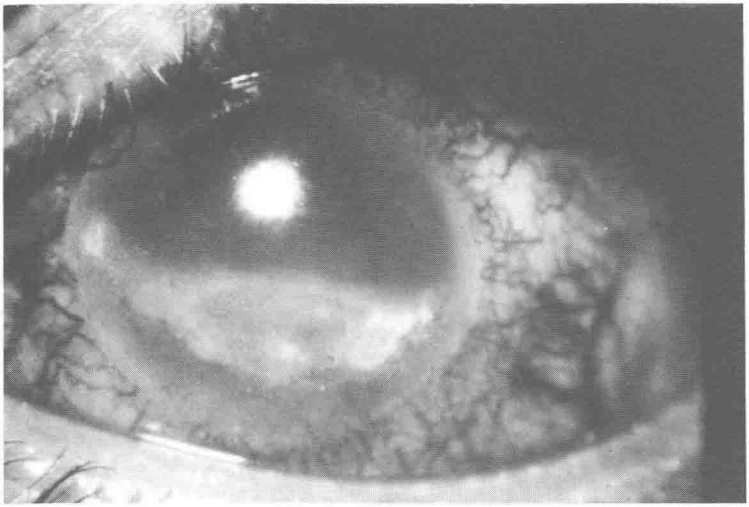

Figure 8 Severe band keratopathy.
(Dr Henry Kaplan.)

Under the slit lamp microscope the cornea has a finely spotted appearance known as punctate epithelial keratopathy. The dull grey spots are tiny patches where the epithelium is devitalized or absent. Dyes such as fluorescein or Rose Bengal adhere to these areas and make them easier to see. Similar spots occur on the conjunctiva but are difficult to detect without the contrast of Rose Bengal staining.

Rose Bengal appears a very dark blue under green light. A small drop is inserted in the lower fornix of the conjunctival sac, for example on the end of a sterile glass rod. It usually stings and provokes watering of the eyes. The epithelium is examined after the patient has blinked once or twice. Patients need to be able to wash the dye from their eyelids before leaving the clinic, and care must be taken not to get it on their clothes. It is a simple test to perform but unfortunately the need for magnification to see the spots in typical cases makes it difficult for the internist to detect all but gross cases with his unaided eye.

3 Keratitis

Various forms of active inflammation of the cornea may accompany systemic diseases. The affected cornea is hazy, and the eye is red. Pain and watering are usually marked. An epithelial keratitis is typical of acute Herpes simplex lesions of the eye (q.v.). However, keratitis associated with systemic disease is more likely to involve the stroma of