Ocular Cicatricial Pemphigoid

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by C. Stephen Foster, M.D., F.A.C.S. and Saadia Rashid
Ocular Immunology and Uveitis Service
Massachusetts Eye and Ear Infirmary
Harvard Medical School
Boston, MA, USA

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Introduction

The term “cicatricial” means “scar” in Latin while “pemphigoid” means “blister” or “swelling” in Greek. Cicatricial pemphigoid (CP) is a systemic autoimmune inflammatory disease that primarily affects the part of the eye called the conjunctiva, a mucous membrane lining the outside of the eyeball. It is part of a spectrum of disorders termed mucous membrane pemphigoid (MMP) that affects other parts of the body, including the skin and mucous membranes lining the mouth, esophagus, trachea, nose, vagina and rectum. MMP and its various subsets are caused by antibodies directed against an otherwise normal part of the body; this results in inflammation and damage to the affected tissue. The average age of onset of CP is 60 to 70 years but patients in their thirties have been reported with this disease. Females are affected more than males but there are no differences in geographical location or race. It is difficult to estimate how common the disease is owing to the difficulty of early diagnosis, but as many as 1 in 8000 individuals may develop CP each year.

History

CP has existed since ancient times. The word pemphigoid was first used by Hippocrates. Wichmann first described CP affecting the conjunctiva in the 18th century. Since then, many scientists have contributed to the study of this disease, referring to it by various names. The name ocular cicatricial pemphigoid, or OCP, is commonly used today, but because of the additional systemic manifestations, CP or MMP is the preferred name.
Course of Disease

Patients may or may not have more than one site affected by the disease. However, 70% of the patients with CP have eye involvement. At the onset (stage I), only one eye is typically affected. Symptoms of conjunctivitis develop: irritation, burning, tearing, mucoid discharge and red eye (Figure 1). These symptoms usually continue, but may stop. Eventually, the condition worsens with development (usually within 3 to 4 years) of a similar conjunctivitis in the other eye.

The disease progresses to conjunctival scarring and shrinkage (stage II), (Figure 2). As the disease worsens, adhesions called symblepheron form (Stage III) (Figure 3). In the end (Stage IV), untreated CP progresses to profound dry eyes, inability to move the eye due to immobilization from ankyloblepharon (adhesions forming between the upper and the lower eyelids) and blindness of both the eyes due to corneal scarring (Figure 4). It may take the untreated disease 10 to 20 years or more to reach the end stage, with bilateral blindness the result. The more advanced the disease, the more likely it is to progress significantly within the next 2 years.

Diagnosis and Testing

A thorough history and eye and systemic examination are essential for a working diagnosis, which is then confirmed by conjunctival biopsy. 83% of cases of CP are correctly diagnosed with conjunctival biopsy. The remaining 17% have a negative biopsy but clinical characteristics are so typical of CP that the clinician proceeds with therapy despite the negative biopsy. Certain eye infections, trauma, topical eye medications taken for some other purpose, and systemic diseases, e.g. sarcoidosis, can also cause conjunctival scarring. Hence, the treating physician may need to perform blood tests and other diagnostic tests, in addition to the conjunctival biopsy to reach a diagnosis.
Cause of Condition
CP is an autoimmune disease, in which body’s own white blood cells, meant to protect the body against germs, start attacking parts of patient’s own body, making antibody against it.

Treatment
CP requires systemic immunosuppressive/immunoregulatory therapy. The medications suppress the patient’s “rebellious” white blood cells to stop the self-destruction of body tissues. They are more effective and much safer than previously used systemic corticosteroids, if prescribed by a chemotherapy expert. Eye drops alone are not beneficial. Symptomatic treatment is also usually required for complications of CP. Dry eyes are treated with ointment lubricants, artificial tears and tear conservation measures. Lid hygiene and oral antibiotics are recommended for lid infection and inflammation. Removal of ingrowing eyelashes is essential to prevent them from damaging the cornea. Other measures such as surgical correction of eyelid deformities, are more successful when the disease has been under control for an extended time.

Prognosis
Permanent remission is usually possible if the disease is diagnosed early and treated sufficiently for 1 to 5 years. The purpose of long-term treatment is to put a stop to the self-destructive autoimmune process. Failure to do so results in invariable progression of the disease, culminating in progressive scarring and bilateral blindness.

Research and Future Outlook
Current research on CP shows promise. The gene that predisposes an individual to develop “rebellious” white blood cells has been identified, as has the target antigen that comes under attack. This information is currently being used in research to develop better diagnostic and therapeutic interventions.