



Figure 1 – A blister has developed on the roof of the mouth in this patient. Such blisters are commonly seen in pemphigoid, although they break rather quickly in most cases.

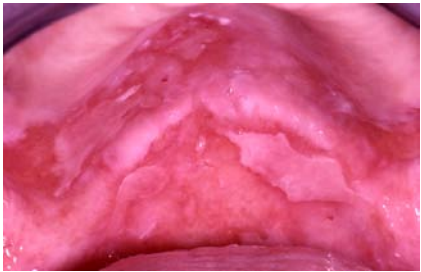


Figure 2 – The roof of the mouth of this patient shows several sores as a result of blisters in this area breaking.

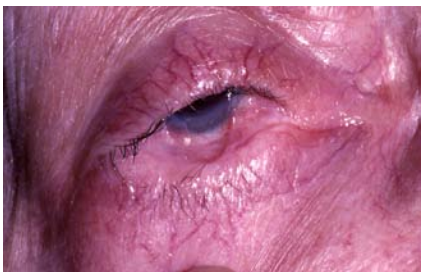
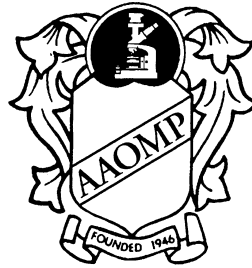


Figure 3 – While involvement of the eyes is not common, it happens occasionally. Patients must be seen regularly by an ophthalmologist (physician specializing in eye diseases) in case the eyes should become affected. Early recognition and treatment can usually prevent blindness.



DIAGNOSIS • TREATMENT
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Cicatricial Pemphigoid



What is cicatricial pemphigoid (sic-uh-tri-shul pem-fuh-goyd)?

Cicatricial pemphigoid is a relatively uncommon blistering disease that mostly affects the mucous membranes, that is, the moist linings of the mouth, the eyes, the nose and throat, and the vagina. Sometimes only one or two of these areas are affected at first, and the condition may spread to other areas, including the skin, if it is not treated.

Who gets cicatricial pemphigoid?

Cicatricial pemphigoid is a condition that primarily affects middle-aged or older adults. Either sex can get the disease, but women are affected slightly more often than men.

Can I spread this disease to my family and friends?

No. Cicatricial pemphigoid is not a disease that can be passed from one person to another like the flu. Probably the best explanation as to what is happening in cicatricial pemphigoid is that it is a type of unusual allergic reaction. In the case of cicatricial pemphigoid, however, instead of being allergic to, say, strawberries or seafood, the body is sort of allergic to itself. In other words, the immune system, which normally protects the body by destroying invading organisms, gets confused and actually starts attacking the lining tissues of the body itself. Thus, cicatricial pemphigoid is in the group of diseases that we call autoimmune (literally “self” immune) diseases.

How do doctors diagnose pemphigoid?

The best way to diagnose pemphigoid is to take a tissue sample (biopsy) from the involved area of the mouth. This is a minor surgical procedure, performed using local anesthesia (numbing) in the office. The tissue is then examined in the laboratory under a microscope and special tests are performed to detect the abnormal immune reaction.

Why is it important to diagnose pemphigoid?

The most serious problem associated with untreated cicatricial pemphigoid is possible blindness if the lining of the eye is involved. The word “cicatricial” means “scarring”, and if the disease affects the eye, the scarring that results often leads to blindness if the condition is not diagnosed early enough and treated properly. Not everyone who has pemphigoid of the mouth will develop pemphigoid in their eyes, but about one person in four will. This is why we advise our patients to be evaluated by an ophthalmologist (medical doctor specializing in eye disease) who is familiar with the signs of eye involvement with cicatricial pemphigoid.

How is pemphigoid treated?

The type of treatment for cicatricial pemphigoid usually depends on the extent of the disease. There are several ways to treat pemphigoid, including tetracycline with niacinamide, cortisone-type drugs that

suppress the immune reaction, or a sulfa-type drug called dapsons. With mild oral involvement, topical (surface) application of a cortisone-type medication a few times each day may be able to control the symptoms. Another alternative is the use of tetracycline and niacinamide, taking one capsule of each, four times daily. On the other hand, if symptoms are more generalized or if the eye is involved, then more serious, cortisone-type drugs that are taken internally may be necessary. If the cortisone-type drugs don't work well or if their side effects are too severe, then dapsons may be tried. A special blood test has to be done, however, to find out if the patient can take dapsons safely.

Can pemphigoid be cured?

In the sense that strep throat, for example, can be cured by a shot of penicillin, no, pemphigoid cannot be cured. The symptoms, however, can usually be controlled with the right medication or combination of medications. Pemphigoid is a condition that may wax and wane on its own to a certain extent, but it usually doesn't go away completely. Fortunately, it rarely causes death, but the possibility of blindness is certainly serious, and the sores and blisters can be quite a nuisance. The goal of your treatment, then, is to keep the sores and blisters under control so that you can lead a relatively normal life. But please realize that this is a very difficult and complex disease to manage, and cooperation and communication between the patient and doctor are necessary to achieve the best possible treatment results.