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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
The oral mucosa is folded, white, and looks
like the tongue, and salivary glands.

Lips, on the other hand, is
since, microscopically, both of
coming in situ, the terms are
from 3% to 5% of squamous
a white patch. Squamous cell
ul in a later chapter (p. 461).
us spongiosis albus mucosae;
tis; congenital leukokeratosis
vesus is a hereditary or familial
or it may appear in childhood,
imum severity in adolescence.
ages occur during the patient's
of a patient's family may have
thickened, and parboiled and

is soft and spongy to touch (Fig. 13-10). The surface may be
folded and may show areas of desquamation. The lesion,
which is asymptomatic, may involve the entire oral mucosa
or be distributed in patches, and it may be associated with
similar lesions on the vaginal and rectal mucosae. The cheek
mucosa is always involved and the gingival margin almost
never.

Microscopically, the epithelium is thickened and shows
acanthosis, but the most striking feature is that the epithelial
cells fail to take any stain and have a "washed-out" appearance
(Fig. 13-11). The connective tissue underneath the epidermis
may show plasma cells and lymphocytes. The lesion
is benign and should be left untreated.

Hereditary benign intraepithelial dyskeratosis (red eye).
Very similar to the white sponge nevus, hereditary benign
intraepithelial dyskeratosis represents a mutation that has

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