



Pachyonychia Congenita Project

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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.

KERATOSIS FOLLICULARIS WITH CHANGES IN THE SKIN,
MUCOUS MEMBRANE AND NAILS

Report of a Case

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We recently observed a case of keratosis follicularis with changes in the oral mucosa and finger nails in addition to the usual cutaneous changes. A review of the current literature shows that such a combination is not common. Ormsby and Montgomery¹ mentioned that nails show subungual hyperkeratosis and changes in the nail plate, and they referred to Reenstierna,² who described lesions on the tongue. Sutton and Sutton³ referred to Brünauer⁴ and Frost⁵; Frost described the occurrence on the gums, at the level of the bicuspsids and molars and extending to the hard palate, of minute, whitish papules, which give a rough sensation to the touch.

Our patient was a woman aged 20, who worked in a printing office wrapping paper. She had noticed her dermatologic disorder for five years and stated the belief that the spongy, hypertrophic gums had been present since the advent of her permanent teeth. At times, the gums bled when the teeth were cleaned; the nails had been easily broken and had split at the distal margins for about four years.

The abnormality of the skin was made up of small, firm, reddish papules, which had been discrete at times but had become confluent, especially in the supraclavicular and infraclavicular regions and in the hair line on the anterior temporal region on both sides. These papules were greasy to the touch. There were a few papules, more nearly the normal color of the skin, around the nasolabial folds. On removal of some of the crusted, confluent papules in the clavicular region, a red, follicular, enlarged opening was seen. The gum of the anterior portion of the upper jaw was hypertrophic to such an extent as to interfere with speech. This hypertrophy was red and spongy to the touch. There was no pain on pressure, and the gums of the lower jaw and the posterior portion of the upper jaw were normal. There were no lesions elsewhere in the mouth. The finger nails were thin and longitudinally ridged and were broken irregularly and split longitudinally at the distal margins, with characteristic changes.

From the Department of Dermatology, Loyola University School of Medicine.

1. Ormsby, O. S., and Montgomery, H.: *Diseases of the Skin*, ed. 6, Philadelphia, Lea & Febiger, 1943, pp. 508-512.
2. Reenstierna, J.: *Dariersche Dermatose mit Schleimhautveränderungen und impetigoartigen Eruptionen*, Arch. f. Dermat. u. Syph. **124**:841, 1918.
3. Sutton, R. L., and Sutton, R. L., Jr.: *Diseases of the Skin*, ed. 9, St. Louis, C. V. Mosby Company, 1935, pp. 592-597.
4. Brünauer, S. R.: *Ueber Schleimhautveränderungen bei Morbus Darier and über die Pathogenese dieser Erkrankung*, Acta dermat.-venereol. **6**:131, 1925.
5. Frost, K.: *Dyskeratosis Follicularis (Darier's Disease)*, Arch. Dermat. & Syph. **31**:508 (April) 1935.

Histologic examination of a portion of skin removed from the clavicular region and stained with hematoxylin and eosin showed acanthosis and disruption, irregular in outline, of the basal layers. There were round bodies in regions of dyskeratosis and grains in the adjacent areas of parakeratosis. These changes took place in the region of the opening of sebaceous glands on the skin.

At the time of this report, the patient has been under treatment for four months, with considerable improvement. She has taken 200,000 units of vitamin A daily for twenty days and has received several grenz ray exposures, as Wise⁶ suggested. The involved skin has become smooth, and the single lesions have disappeared. The hypertrophic gums have receded, and the patient states they no longer bleed when brushed. The finger nails do not split as they formerly did. Instead of the hard, oily, seemingly crusted skin of the clavicular regions, the area is smoother and nonscaly. Residual brown pigmentation is present.

SUMMARY

A case of keratosis follicularis is reported because of the unusual changes in the mucous membrane and in the finger nails, as well as the usual changes in the skin.

Vitamin A therapy produced a pronounced amelioration in the size of the cutaneous lesions. Grenz rays were also used and believed to have been of benefit.

706 South Wolcott Street.

6. Wise, F., in discussion on Bechet, P. E.: Darier's Disease Associated with Possible Diabetes Mellitus, Arch. Dermat. & Syph. 35:1205 (June) 1937.

REACTIONS TO SODIUM BISMUTH TRIGLYCOLLAMATE ("BISTRIMATE")

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Recently there was a report¹ on the efficiency of sodium bismuth triglycollamate tablets ("bistriplate tablets," Carroll Dunham Smith Pharmacal Company) for the treatment of lupus erythematosus and other chronic dermatoses.

To date the only report of untoward reactions from use of the drug which has come to our attention is that made by Gross,² of temporary gastrointestinal disturbances, with anorexia, nausea and vomiting. During the past months we treated 2 patients, 1 of whom had gastric disturbance in addition to oral lesions, and the other an entirely different series of reactions involving the skin and mucous membranes, following ingestion of "bistriplate."

1. Sulzberger, M. B.: 1945 Year Book of Dermatology and Syphilology, Chicago, The Year Book Publishers, Inc., 1946, p. 31.
2. Gross, E. R., in discussion on Wright, C., and Gross, E. R.: Disseminated Lupus Erythematosus Treated with Sodium Bismuth Triglycollamate ("Bistriplate Tablets"), Arch. Dermat. & Syph. 55:862 (June) 1947.