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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.
I diluted it with the patient's own serum and found it made no difference in the cutaneous reactions which could be elicited. I am familiar with the work of D. S. Martin and D. T. Smith at Duke University, who reported a series of cases of blastomycosis (Blastomycosis: A Review of the Literature, Am. Rev. Tuberc. 39: 275 [March] 1939; A Report of Thirteen New Cases, ibid. 39:488 [April] 1939). They found that after desensitizing the skin to blastomycin, the therapeutic response was better to iodide therapy. They also mentioned that it might be dangerous to give iodides to such patients without first desensitizing the skin to blastomycin. This patient was given iodides in doses as high as 600 grains (39 Gm.) a day and over long periods without any evidence of exacerbation of the lesions. While I was successful in curing an area by curettage followed by application of gentian violet, the surgeons refused to undertake extensive curettage under anesthesia. At the present time the patient has just completed two weeks' treatment with thymol, and it is of interest that he was able to tolerate a dose of 4 Gm. a day without symptoms. The usual dose is from 0.5 to 2 Gm. a day. There seems to be more improvement from thymol than from any other form of therapy which he has had.

**Pachyonychia Congenita.** Presented by Dr. Anthony C. Cipollaro, New York.

J. P., an American boy aged 12 years, is presented from the Skin and Cancer Unit of the New York Post-Graduate Medical School and Hospital. His mother says that the patient was born with deformed nails. The changes in the oral mucous membranes and the skin occurred soon after birth and have gradually increased in severity.

Examination shows on the inner surfaces of the lips and cheeks and on the dorsum of the edges of the tongue, dull whitish gray patches, irregular in shape and slightly elevated. In some places the patches have an irregular surface, somewhat suggestive of a verrucous condition. These leukokeratotic patches show no signs of malignant degeneration. The nails of both hands and both feet are abnormal. They are yellowish brown, distorted and hyperkeratotic. The nails are hard and oversized, as in cases of onychogryphosis. The skin shows the following changes: On the mesial surfaces of both great toes are areas in which there are sharply demarcated patches of denuded skin. These areas are glistening and show little tendency to heal and are gradually progressing peripherally. On the outer side of the right foot, just below the external malleolus, there is a similar patch measuring approximately 7 by 3 cm. The skin as a whole is dry. On the external surfaces of both arms, there are many fine keratotic lesions. Verrucae of varying sizes are present on the tips of the elbows and the extensors of the forearms.

The Wassermann and Kahn reactions were negative. Cultures from the tongue for fungi were negative.

Treatment has consisted of topical applications.

**DISCUSSION**

Dr. A. Benson Cannon, New York: I agree with the diagnosis in this case. I should like to call attention to the lesions of the mucous membranes that so closely resemble the condition that I described a few years ago as white sponge nevus of the mucous membranes of the oral cavity, vaginal mucous membranes and anus (Cannon, A. B.: White Sponge Nevus of the Mucosa [Naevus Spongiosus Albus Mucosae], Arch. Dermat. & Syph. 31:365 [March] 1935). The only difference that I can note in this patient's lesions of the mouth from the condition in my patients is that they have sialolike, clear, hyaline, discrete, raised, pinhead-sized areas in the white spongy parts that resembled vesicles. Their disease was congenital and appeared in three different generations of the same family. Since publication of the history of those cases, I have studied two
other family groups with the same condition, no member of either group having any other defect.

Dr. Donald M. Pillsbury, Philadelphia: I agree with the diagnosis as presented. In connection with this case, I recall a report by Goldberg (Goldberg, S. C.: Resistant Erosive Lesions in Pachyonychia Congenita of Jadassohn: Treatment with Buffered Cysteine Hydrochloride, Arch. Dermat. & Syph. 36:331 [Aug.] 1937), in which ulcerated lesions in a case of this disease healed after applications of cysteine hydrochloride to promote epithelization.

Dr. George C. Andrews, New York: There are reasons to believe that this condition is a variant of Darier's disease, keratosis follicularis. Papules develop on the extremities which are indistinguishable from the lesions of Darier's disease.

**Naevus Cerebriformis (Cerebelliformis) of the Scalp.** Presented by Dr. Ellwood C. Weiss, Bridgeport, Conn.

H. F., a woman aged 24, a private patient, is presented with a large nevoid process involving the scalp since birth. On the scalp, beginning about 9 cm. back of the front hair line and extending transversely over the scalp from ear to ear and from nose to neck, involving the entire vertex, occipital, and suboccipital regions, is a large pinkish, irregularly convoluted mass, 25 by 25 cm., practically devoid of hair except from the depths of the sulci between the convolutions where a fair amount of hair arises to provide a partial covering for the tumor mass. The convolutions are irregular and unusually large, as evidenced by the lowermost transverse convolution in the suboccipital region, which hangs down in an almost apron-like effect and measures 25 cm. from side to side, its width being 4 cm. and its depth over 5 cm. The borders of the lesion are almost precipitously from what appears to be normal skin of the scalp anteriorly and normal skin of the neck posteriorly and at the sides as well. The patient's mentality is average or above. There is a somewhat foul odor present. There is no familial history of similar tumors. The patient's daughter, however, presents a small naevus flammeus of the left forearm.

The histologic report was as follows: The sections of a cutaneous tumor showed atrophic epidermis. A moderate hyperkeratosis was present. Large sebaceous glands could be seen. The stroma of the tumor showed marked fibrosis. It contained numerous dilated thin-walled vessels. In the upper portions of the stroma were numerous polygonal cells arranged in nests. These were typical nevus cells. Here and there could be seen pigment-containing cells. A few chromatophores were noted. Mast cells were scattered throughout the section. The deeper portions of the growth showed connective tissue which contained numerous bundles of spindle-shaped cells. Here the growth suggested the picture sometimes interpreted as a neurinoma. Most of the cells, however, appeared to be spindle-shaped nevus cells. The histologic diagnosis was nevus pigmentosus (Dr. Gerald F. Machacke).

**DISCUSSION**

Dr. Ellwood C. Weiss, Bridgeport, Conn.: This case was presented principally for two reasons. One was because of the large size of the nevus and the other was for suggestions as to treatment. The case was primarily that of a surgeon and was referred only recently to me for an opinion. I have seen only a few cerebriform nevi which might approach the size of this one. Aside from the size of the lesion and its appearance, there is an odor from it, and it is objectionable to the patient on this account. She is only 24 years old and is anxious to have the lesion removed. The question in the mind of the surgeon is whether to remove it in one stage, followed by skin grafting, or to do so in several stages, performing a partial excision of one quarter of the lesion at one time, then having it healed together for summer if the tissues permit and eventually removing the entire lesion in stages in this fashion. It is surprising how little one can find in the dermatologic textbooks or literature on this subject.

It is hard to say whether a similar subject might not occur in the nevus of Jadassohn; hence, the stages.

Dr. Donald M. Pillsbury, Chicago: The dermal lesion can be managed only by excision of nevus of Jadassohn. The growth is not, and the patient should understand that.

Dr. James Filmore: The problem is comparable to that of a man whose presentation is to determine the choice of treatment. I should think that in the early stages of healing, it is better, if skin grafting is contemplated, to have the only raw area covered.

Dr. Minor: We have had no such case, but I think that this patient's treatment is better than this patient's opinion.

Dr. Albert Von Lengyel: We find the nevus of Jadassohn is not a nevus of any great size, except when locally with the jaw, it becomes a large one.