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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.
DR. FLOMA: We were able to obtain the names of 3 contacts, 1 of whom has latent syphilis, and we are still looking for the other 2. Mixed infections are not unusual. About 3% of our patients with gonorrhea also have infectious syphilis. We have had several cases this year of mixed infections, chancroid and secondary syphilis, chancroid and primary syphilis, gonorrhea and chancroid. One of our patients had gonorrhea and granuloma inguinale.

Pachyonychia Congenita. Presented by Dr. CLEVELAND R. DENTON.

A female infant aged 18 months, of Irish and Scottish ancestry, has had a lifelong dermatosis involving the upper and lower extremities, nails, and mouth. The patient’s nails were normal at birth, but within 3 or 4 days they became white and fell off. Abnormal nail growth occurred and has continued. The patient is reported to have had a white dermatitis of the mouth, which was treated by antibiotics and gentian violet and cleared within 1 month. At the age of 1 year she developed dry, erythematous papules about the knees when she crept. Bullous lesions about the toes occurred when the child started to walk 4 or 5 months ago.

The nails of the fingers and toes are hypertrophic, and there is slight hyperkeratosis of the palms and soles. White, hyperkeratotic lesions are seen on the tongue, anterior buccal mucosa, and anterior portion of the hard palate. There are bullous lesions of the feet and hyperkeratotic lesions on the elbows and knees.

No treatment was given until recently, when cool foot-soaks of potassium permanganate, 1:12,000, were prescribed, with Cortisporin* ointment locally.

Pachyonychia Congenita. Presented by Dr. CLEVELAND R. DENTON.

A 16-year-old girl, sister of the preceding patient, is presented from Hartford Hospital with a lifelong dermatosis involving the nails, hands, feet, extensor aspects of the extremities, and the mouth. The patient has had hyperkeratosis of the palms and soles, with blister formation, and hypertrophic nails since birth. Her mother and a maternal aunt aged 16, as well as her infant sister, have a similar condition. The maternal grandmother had 2 hypertrophic fingernails, and a maternal uncle who died of pneumonia had hypertrophic nails. The patient incidentally has epilepsy.

The nails are hypertrophied, and the patient has a generally dry skin, with hyperkeratotic and follicular papules over the extremities, particularly about the extensor aspects of the elbows and knees. There is marked hyperkeratosis of the hands and feet, with fissuring of interdigital webs of the feet, as well as ruptured bullous lesions of the soles.

* Polymyxin B sulfate, bacitracin, neomycin sulfate, and hydrocortisone.
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white, hypertrophic lesions on the back of the hand, buccal mucosa. The gums are hypertrophic.

A gross examination of skin scrapings from the toes was negative for mycelia and sporulation was cultured for fungi, but not available for reading as yet. Direct immersion of oral lesions was negative of the same material yielded a growth of a type not so far determined.

1 Dermatological therapy has been pre

The foot. Diphendylhydantoin (Dilantin) is useful in epilepsy, which probably accounts for reddened gums. A course of vitaminated 1 week ago, 100,000 units to be continued on a week off.

nia Congenita. Presented by Dr. R. R. DENTON.

aged 35, of Scottish and Irish ancestry, is one of the 2 preceding patients. She has an interesting condition of hypertrophic nails; a warty, papular dermatitis of the elbows with a generally dry skin; white, hyp

nails of the tongue and anterior aspects of mucosa and hard palate; bullous intact and ruptured, of the soles, with heeled feet which had received no treatment until she advised use of cold potassium permanganate and Cortisporin ointment locally.

Discussion

LE KIRSCH: At the New York Skin Unit, Dr. John Garb presented a mother of the same type of process, including atosis oris and the great amount of sores of the soles and the thinned nails. She received special shoes so these people around, and they did fairly well. The report in the Archives of Dermatology 1959 (62:117 July 1950) showing the shoe and the follow-up studies, and it what relief they had. Practically all atosis disappeared. The shoes are very good, you have to keep making new ones.

RO. S. YAFFEE: One intriguing thing about this is that these feet form bullae on their feet if it is so, which is divided into cystine space lesions. The most unusual there was a report by Fred Mullins from Texas resulted in amelioration of pain in 3 days and abatement of pain in 2 weeks his patient, a boy, walked with a reasonably normal gait and apparently could tolerate shoes for the first time. The hyperkeratosis was reportedly diminished. It is astounding that congenital hyperkeratoses or keratotic dystrophies could be influenced by hypnosis. The most celebrated case was that of Mason, a patient with congenital ichthyosiform erythroderma who was improved by hypnosis. It was reported to the British Medical Association in several publications.

DR. HARVEY B. ANSELL: During hot weather sufferers with this disease may be confined to bed because of severe pain on walking. In an attempt to relieve the pain of a patient with a similar condition I prescribed steroids in relatively small dosage. The results were dramatic. When the steroids were discontinued the condition relapsed.

DR. HOWARD S. YAFFEE: Space shoes are not necessarily extremely expensive. In the less expensive models a plastic mold is made and kept on file. New pressings may be made from the impression at a later date.

DR. CLEVELAND R. DENTON: Recently, William Buckley et al. mentioned that pachyonychia congenita seldom continues over more than 2 generations. In this instance 3 generations are involved, beginning with the maternal grandmother and extending through the mother and daughters, who had virtually all the reported abnormalities seen in this disease.

Nutritional Cirrhosis with Purpura and Hypergammaglobulinemia ("Secondary Purpura Hypergammaglobulinemidae"). Presented by Dr. HERBERT MESCON.

A woman aged 51, a Civil Service employee, is presented from Lebanon Shattuck Hospital with an 8-year history of recurrent purpuric eruptions on the legs. The patient has documented nutritional cirrhosis, emphysema, and mild diabetes. The eruptions are characterized by (1) recurrent showers of petechiae with gradual confluence, darkening, and residual hyperpigmentation; (2) transient burning and sensations of tenderness in the legs at onset; pruritus is unusual, but there is occasional ankle edema; (3) precipitation by prolonged dependency of the extremities; (4) recurrence every few weeks in the past several years, occasionally involving the thighs, lower part of the abdomen, and forearms.

There have been mild to moderate episodes of ascites in recent years but no significant jaundice or gastrointestinal bleeding. Varicosities developed in the lower extremities in 20 years ago, with periodic edema of the ankles since then. There have been no stasis ulcers. High saphenous vein ligation was performed 16 years ago. Three years ago thrombophlebitis of the lower extremities occurred.

The patient shows cyanotic lips and nail beds, with clubbed fingers. Hepatosplenomegaly, mild ascites, and edema of the legs are present. The peripheral pulses are adequate, and no jaundice is noted. Varicosities are seen on the thighs.

The skin shows new and old petechiae and hyperpigmentation in discrete and confluent, reticulated patterns, with scaling on the legs to the level of the knees. An eschar is present on the left lateral malleolus area. There are scattered, fading petechiae on the lower, anteromedial aspects of the thighs, and irregular hyperpigmentation on the extensor aspects of the forearms. Facial telangiectases and spider angiomas are present.

Laboratory Findings.—The serum iron and iron-binding capacity were normal; nonprotein nitrogen, 30. Urinalyses were normal except for a trace of 2+ sugar. Two hours after eating the blood sugar was 114. Bone marrow studies were normal. A tourniquet test performed on the right forearm an April 23 was positive. An upper gastrointestinal series was negative for esophagial varices. White blood cells numbered 3,000, with a normal differential count. Serum transaminase (SGOT) was 46; bleeding and clotting times were normal; SBF, 34% retention in 45 minutes; prothrombin time, 82% of normal; bilirubin, 1.1; hemocrit, 44; ESR, 18. Total proteins equalled 9.1 (albumin 3.8, globulin 5.3). Serum electrophoresis was nonspecific, marked increase in γ-globulin (3.64 g/dl; normal, 0.6-1.3).

Treatment has consisted of diated baths; a high-protein, low-sodium diet; periodic use of diuretics; tolbutamide (Orinase), 0.5 g. bid; and periodic use of bromocriptin.

Discussion

DR. HERBERT MESCON: The first impression, at least on looking at the legs, was that this is a stasis purpura or perhaps even one of the pigmented purpuric eruptions. On the other hand, she did have purpuric lesions on her arms, as well as on the legs. If we do not think in terms of hypergammaglobulinemia as a possible cause of purpura, then we will not make the diagnosis.

In this case, we feel it is not idiopathic, but secondary to the hepatic disease, and that was confirmed by evidence of liver disease as well as by the electrophoretic pattern.

How hypergammaglobulinemia results in petechiae is not well understood. One explanation is that the abnormal globulins are increased, and abnormal γ-globulins somehow affect the vessel walls so that they are incapable of withstanding the usual types of stress. Considerable work in animals has shown that the ability of the blood vessels to withstand rupture is altered by such things as nutritional deficiencies, not only of vitamin C, but even fat. Endocrinopathies will interfere with the ability of the capillaries to withstand pressure.

Wiener recently presented a paper at the American Dermatological Association meeting on purpura...