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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.
bodies were found in an intra- and extracellular distribution. These bodies were nonencapsulated, round to oval, and were approximately 2-4 μ in diameter (Lever, W. F.: Histopathology of the Skin, Ed. 2, J. B. Lippincott Company, 1954, p. 237), and they did stain a very dark red with Giersa's stain. The parasun could not be identified in the examined organisms. I believe the parasun can be better visualized on recovering the organisms after culture. Subsequent specimens from the oral pharynx also revealed a granulomatosus infiltrate and the identification of Leishman bodies. Therefore, this patient has mucocutaneous or American leishmaniasis (espana).

Dr. Murray Zimmerman: Thank you, Dr. Hirsch. Leishmaniasis is what I was suspecting was being held out on us. I think Miss Avis Gregersen thought so, too, since the first thing she asked for was a Giensa stain.

Dr. Muller, would you like to elaborate briefly on what the Montenegro skin test is?

Dr. Sigfried Muller: This patient comes from the Choco area in Colombia where leishmaniasis is endemic. He recently spent much time in the forest, sleeping outdoors without protection, and this is usually a requirement for developing leishmaniasis. Some confusion exists about the occurrence of the mucocutaneous manifestation in the American leishmaniasis. This usually occurs several months or years after the initial cutaneous lesions, and often it never occurs. In Panama the mucocutaneous complications are seen in less than 2% of patients who develop leishmaniasis. In a series of over 70 patients that I studied in Panama, over a 2-year period, one-fourth of them had only a solitary ulcer, usually on the exposed areas. The most important point in making a diagnosis of leishmaniasis is to think of it. Some useful procedures for making the diagnosis are the following: (1) the Montenegro skin test, which has a similar significance to the tuberculin skin test; (2) identification of the parasite in smears of serum obtained from the undermined edge of the ulcer, or identification in histopathologic sections; (3) culture. The leptonoma phase is seen in culture and, in experienced hands, can be very valuable. Disseminated Granuloma Annulare. Presented by Dr. Max Popenoe. Trichophyton Rubrum Tinea Pedia, Recurrent After Griseofulvin Therapy. Presented by Dr. STANTON B. MAY. Melanoma in a Pigmented Nevis Treated with Radiation. Presented by Dr. STANTON B. MAY. Granulomatous Ankle Ulcer Culturing Proteus Organism. Presented by Dr. WILLARD MARMELAT. Recurrent Peritonsillitis in Pregnancy Due to Gonococcus. Presented by Dr. STANTON B. MAY.

SOCIETY TRANSACTIONS

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Dr. Alford C. Fishman: This is the first case of such a patient that I have ever seen, but I recall what Dr. Hyman in teaching us paronychia congenita there was. Now, where would one biopsy,
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Dr. Alford C. Fishman: I feel this in particular case is the disease which I feel, is part of the picture of bullous lesions.

Dr. Jack E. McCreary: In this particular case is the disease which I feel, is part of the picture of bullous lesions.

In our office practice we find it interesting to observe a Negro family, older 2 of her 3 children with paronychia congenita. This family shows at times, marked plantar areas, and acanthasis follicular keratoses, changes of the disease. We have seen this phenomenon at our own institution. The keratoses have continued to improve during pregnancy and menstruation. The keratoses diminished as she has grown. She also stated that she had 5 months without benefit.
over pressure areas of the feet, this presentation, I feel, is part of the picture of epidermolysis bullosa. In fact, severe plantar hyperkeratosis over pressure areas may well be a variant of epidermolysis bullosa. I have a patient under observation at present who has severe plantar calluses and also corns over pressure and friction areas. She gave a history that in the past she developed blisters on the feet, hands, and knees after trauma; at present, however, the traumatic blisters are insignificant. It may be that she represented a form of epidermolysis bullosa. For treatment, I should suggest the use of contour shoes which should at least improve the tendency to form calluses. You may recall the article by John Garb (Pachyonychia Congenita: Regression of Plantar Lesions on Patients Wearing Specially Made Leather Base Foot Mold and Shoes, Arch Derm Syph 62:117—124 [July] 1950), in which severe plantar calluses in a mother and son were dramatically improved by wearing the special shoes. Garb’s patients also presented the combination of severe callousness and blisters.

**Dr. Harold C. Fishman:** I must admit that this is the first case of pachyonychia congenita I have ever seen, but I remember Dr. Arthur Hyman in teaching us pachyonychia congenita there is a subepidermal bulla. Now would one biopsy to find it? As far as I can tell, there are only 2 involved regions: the nail lesions and, possibly, the hyperkeratotic lesions of the feet.

**Dr. Alfred G. Lerner:** In the condition known as acrodermatitis enteropathica, the patient may have on the extremities vesiculobullous lesions which look like epidermolysis bullosa. The treatment for the former condition is Diodoquin. I wonder if anyone has used Diodoquin in the treatment of epidermolysis bullosa.

**Dr. Jack E. McCleary:** The distressing feature in this particular case is the “blistering” tendency exhibited. Bullae, especially over the plantar surfaces, are reported not uncommonly in pachyonychia congenita, but the development of them over the trunk and extremities is rare.

In our practice we have had an opportunity to observe a Negro family; the mother and the older 2 of her 3 children have pachyonychia congenita. This family showed the typical leukokeratosis, marked plantar and palmar keratoderma, acniform follicular keratoses, and ichthyotic changes of the disease. We have never noted, nor have they described at any time, the occurrence of blisters. The mother believed that her skin problem improved during pregnancy and possibly during menstruation. The keratoderma of the soles has diminished as she has grown older. The mother also stated that she had taken griseofulvin for 5 months without benefit.

**Pachyonychia congenita also has been reported in certain Congo tribes.**

**Dr. Alfred G. Lerner:** In pachyonychia congenita, the patient may show dystrophic changes of the nails, palmar and plantar keratoderma, and leukoplakia of the oral mucosa. A variety of pachyonychia congenita is dyskeratotic congenita. In this condition there are only dystrophic changes of the nails and the absence of leukoplakia; however, there are no hyperkeratotic changes of the skin.

**Dr. Willard Marmelzat:** The one man who claimed some therapeutic success in treating epidermolysis bullosa was the late Dr. John Lamb. In 1956, during the Pacific Dermatology Association meeting in Hawaii, Dr. Lamb described to me his success with the use of pregnenolone acetate. If my recollection is correct, he started with a dosage of 400 mg. of pregnenolone acetate daily for an infant and increased it to as much as 1,000 mg. daily. There were no untoward or adverse effects. The drug is not available here in California but can be obtained in Oklahoma.

**Dr. Rose B. Saperstein:** I’d like to point out that nails like these are seen every day. The mother’s nails certainly look like those in psoriasis or tinea. I think that we often see one nail, usually of the small toe, like that of the child’s, and we would bring up the question, “Is this a forme fruste type of congenital pachyonychia? It seems to me that I have seen many such nails but usually only single nails. This is the first time that I have seen all the nails look like this. Perhaps this will make us all more observant and check more closely to see if the person ever had a bullous formation.

**Dr. Stanton B. May:** I am surprised that someone does not challenge my diagnosis. The nail changes are my concept of pachyonychia congenita. In reading through Pardo-Castello’s book on Diseases of the Nails, I found it difficult to distinguish the various nail abnormalities one from the other. In his discussion of the nail changes in epidermolysis bullosa, he does not mention this change, but speaks of characteristic signs; namely, atrophy, loosening of the free edge, and frequent falling off of the nails. As a very rare change he mentions onychogryposis but no mention of pachyonychia congenita. The mother of the child today called my attention to a blistering of the occiput of the scalp undoubtedly from friction in lying on his back. I know of no treatment, but I do recall when Dr. Belisario of Australia was here he suggested that oral sodium citrate was of value in pachyonychia congenita.

**Melanotic Nevoid Lesion (Benign? or Malignant?),** Presented by Dr. Laurence J. Underwood and Dr. Walter S. Green.

A 43-year-old Caucasian male has had an asymptomatic, pigmented, large flat lesion on his
right lumbar region "all of his life." He states that he has noted no change in any way in the lesion. He consulted the presenters because of varying opinions regarding it.

A deeply pigmented (blackish color) lesion, measuring 3 by 5 cm., is located on the right lumbar region. It is flat with minimal nodularity and very minimal scaliness at the inferior portion. A more lightly pigmented and smaller lesion is located on the medial aspect of the left arm.

**Biopsies:** Specimens were taken from the inferior and superior aspects of the back lesion, and both specimens are adjacent on the single slide.

The unusual color, size, and apparent unchanging characteristics of this "lifelong" lesion, coupled with the histopathologic findings, are of interest. The presenters advised the patient that excision of the lesion is desirable.

**Discussion**

**Dr. Margaret Ann Storkan:** Never before have I seen such a bizarre lesion which has been present a lifetime. At this stage it has, in my opinion, features of anaplasia.

**Dr. Josefa Novaes, Mexico City (by invitation):** I think this is a benign lesion related to a nevus unius lateris in which melanin is also present.

**Dr. Murray C. Zimmerman:** I'd like to ask Dr. Novaes and Dr. Storkan if they could tell what those large clear cells are and if the cells are completely benign.

**Dr. Fred F. Feldman:** I should like an explanation for the cells that are creeping up into the epidermis.

**Dr. Kurt Lindstrom:** Dr. Novaes said that they are perfectly benign.

**Dr. Paul Hirsch:** The sections presented were inadequate for the determination of a definitive diagnosis. I do not think that one should commit himself from the presented sections. I should suggest that the paraffin block of this biopsy be reentered and multiple serial sections be examined. The slides presented did show features of anaplasia of this tumor of melanin-containing cells within the epidermis and upper one-half of the corium. These features of anaplasia were manifested by the transmigration of individual as well as nests of tumor cells throughout the epidermis and ulceration into the epidermal surface, some pleomorphism, and minimal atypism. There were also factors of benignity on the basis of the organized pattern of maturation, relative absence of mitotic figures, and a tendency toward a normal maturation process. Clinically, this was a very large lesion, approximately 7.0 by 5.0 cm., irregular surface, slightly elevated and indurated, black in color, and exhibiting an erythematous or inflammatory halo at its margin.

On the basis of the clinical appearance, the history of its duration, and the variable microscopic picture, I should strongly suggest excision of the lesion in toto with adequate and representative serial sections being studied before a definitive diagnosis be submitted.

It is unfair to request from the attending dermal pathologists a definitive diagnosis of such a significant magnitude on the basis of the material studied as well as the time allowed for its examination. I, therefore, should suggest the above recommendations be performed, and the patient be re-presented at a later date.

**Dr. Murray C. Zimmerman:** Last night, at the L.A. County Hospital staff meeting, a similar case was presented with a question of malignancy or quasi-malignancy. The patient had a lesion which on histologic examination involved the upper one-half of the cutis. Clinically, it was raised, velvety, and strikingly similar in almost every way to that of the patient presented tonight. This was considered as being a malignant melanoma. If these lesions invade only to the level of the sebaceous glands, are they more benign lesions, as has been suggested? Should it be called a Sophie-Spitz melanoma or a superficial melanoma? Should there be a special term for these because they are not as malignant as other melanomas? Again, are they benign?

**Dr. Fred F. Feldman:** Are you referring to juvenile melanoma?

**Dr. Murray C. Zimmerman:** No, I am referring to a superficial type of malignant melanoma which invades and is sharply limited to the upper one-half of the cutis. The one we had last night was different from this one in that it had no round-cell infiltrate. It was just as sharply margined at the upper one-third of the cutis.

**Dr. Stanley Mounce:** I think this is a true melanoma, or lentigo maligna. It's malignant from the beginning. It has all the characteristics of a melanoma. The fact that it hasn't become active up to the present time doesn't mean it won't. It might wait another year or 2 in this phase and then eventually spread very widely. So I feel that, while it is still in the insidious stage, it is a definite malignancy and should be very widely excised and examined more thoroughly. We see lesions in children somewhat like this which are not true melanomas; they are benign lesions, but, in an adult, they wouldn't be called an active junction nevus.

I think Dr. Becker would say that lentigines maligni are melanomas from the beginning. They may remain inactive like this for 10 or 15 years, but all of a sudden they develop into the tumor stage and spread widely. However, the process is malignant from the beginning.

**Dr. Harold Price:** With all due respect to histopathology, it is the clinician who has to make the final decision. In this particular case, the lesion was excised. I suggest a biopsy of a suspected lesion. I believe that we should go by the thought that all lesions of this type are malignant until proven otherwise. I think we should look at them with suspicion, and not just go by a biopsy for all pigmented lesions which we dermatologists easily remove. We have keratoses, pigmented basal-cell carcinomas, which we have seen in many colleagues, and I have seen a documented case of a nevus basilaria which became malignant after incomplete removal. I think we need to be very careful that these lesions are not allowed to become malignant, and we should not allow them to become malignant by incomplete excision.

**Dr. Antis Gelber:** About 1 year ago I saw a young male college student...
the final decision. In this patient, we are dealing with a lesion which was present at birth, a pigmented nevus. In most instances a pigmented nevus in adult life will show little if any junctional activity, but this lesion shows considerable junctional activity and in addition an inflammatory dermal infiltrate. I have a feeling that this is a combined lesion of active junctional nevus and blue nevus; however, there is something about the picture that makes me uneasy, and I would strongly urge complete excision.

Dr. Ralph B. Coomber: On our Tumor Board, some of the surgeons who excise everything except a bathing-suit nevus are horrified when we suggest a biopsy of a suspected melanoma. I wonder, from a medicolegal standpoint, whether any of you have had a similar experience, and what the policy is of other tumor boards.

Dr. William L. Marazzat: I should like to express an opinion on the previous remarks. We are once again justing with the familiar old windmill—is it or is it not safe partially to remove for biopsy purposes a pigmented lesion? I have observed that some problem in many dermatologic societies both here and abroad and have repeatedly heard the identical arguments and counterarguments. There is a great deal of mythology and confusion concerning the management of nevi. There is a popular fear, not only among lay people, but among physicians and surgeons as well, that disturbing a nevus can "start something." Yet all of us who work on tumor boards are familiar with the slow and often contemptuous with which general surgeons view our "less-than-radical" approach to the problem. The surgical literature is dogmatic and will accept nothing less than total excision biopsy for all pigmented lesions (many of which we dermatologists easily recognize as seborrheic keratoses, pigmented basal-cell carcinomas, etc.). I have searched the literature and spoken with many colleagues, and I have yet to find a documented case of a nevus becoming a melanocarcinoma after incomplete removal by any means—high frequency curet, cryotherapy, live cautery, or incomplete excision. We are convinced of the rightness of our position must continue to speak up, sound our opinion, and educate our dermatologic brethren.

Currently I have under my care a young woman with a very large junction nevus on her back (proved benign by punch biopsy). Nearby she has had a basal-cell carcinoma of the back and a second one on the forehead. Would any of you comment or speculate as to the posibility or probability that her active junction nevus is more likely to become malignant because of her known basal-cell carcinoma history?

Dr. Anita Gelber: About 7 or 8 years ago I saw a young male college student who had had for most of his life a flat, brownish-black lesion about the size of a silver quarter on the back of his right leg. The lesion was biopsied, and the question of malignancy was debatable, as various histopathologists differed in their microscopic diagnosis.

Dr. Leo Kaplan studied the sections, and he suggested that even though there were large cells filled with pigment he felt that it was some type of nevus (maybe a borderline one with which the present time we are not familiar). The lesion was removed. The patient has been actively employed as a salesman, and to my knowledge there has been no recurrence.

Dr. Fred E. Feldman: This is a case in point where a small punch biopsy may not be truly representative of the lesion. Our discussion, up to now, has been academic and based on a small sampling of the lesion. When the lesion is completely removed and serially sectioned we may uncover a great deal more inflammatory reaction and possibly more evidence of cellular proliferation into the epidermis. From a clinical standpoint alone, I feel that the lesion should be excised.

Dr. Murray C. Zimmerman: In patients with malignant melanoma, after centrifuging down the blood with silicones, malignant melanoma cells can be isolated from the circulating blood in a high percentage of cases. If you figure these cells are neurogenic and embryologically migrate from the neural crest to the skin, they revert again to their state of almost no cohesiveness. Malignant cells in general tend to lose cohesiveness as they become more anaplastic. At any rate, the people who have malignant melanoma have a routine oncocytoses, or whatever you want to call cancer cells circulating in the blood stream. They can then be given phenylalanine-melphalan. Phenylalanine, of course, is the precursor to dopa (dihydroxyphenylalanine). The malignant cell is doped into picking up this poisonous phenylalanine for its metabolic needs. On doing so it picks up a lethal dose of melphalan at the same time, while other cells that don't use such huge quantities of phenylalanine are not so badly poisoned.

Dr. Laurence J. Underwood: The unchanging features of this lesion, namely, the large size, intense black pigmentation, and lifelong duration, coupled with the very interesting pathologic features, make this case of particular interest. The histogenesis of melanomas has been a study of importance, and for further understanding of the subject the excellent articles of Dr. S. Becker, Sr., and Dr. A. Allen and Dr. S. Spitz in the A.M.A. Archives of Dermatology and Syphilology, Volume 69, 1954, are recommended. Further study of the initial specimen by an expert dermatopathologist was diagnosed as a malignant melanoma. The lesion will be excised by a surgeon.