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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.
noma. The histology varied very much. In some parts there was a rather whorled arrangement suggesting some types of transitional or cell carcinoma. Another nodule removed from the same case suggested, on the whole, a malignant melanoma but of rather different structure from the former nodule, shown from the same case. The whorled arrangement of cells was more marked in the latter.

In a section taken from the third case, there was the same picture but with rather more epithelial hyperplasia, and even epithelial islets deep in the cutis, suggesting the possibility that there might be an epitheliomatous component in addition to the melanoma!

Finally, there was a tumour in which the cells showed a very loose reticular arrangement and included pigmented cells which might indicate a malignant melanoma. Again it was difficult to be certain.

He had shown each of the sections obtained to demonstrate the variety of the histological appearances met with in nodules removed from such cases, and to emphasize the difficulty of forming a definite conclusion as to their nature.

Their clinical behaviour appeared to be much more benign than was usual in actively growing naevus-carcinomas. Their histology was most varied and suggested a greater degree of malignancy than was borne out by their clinical course. The relationship between the different appearances observed in individual nodules was not easily interpreted.

[Postscript.—In the American Journal of Cancer (1938, 33, 196), Stout has called attention to “a group of superficial, slow-growing naevo-carcinomas, with Paget-like characteristics which distinguish them from other melanomas”. He refers to papers by Kreibich, Darier, Civatte and Bloch, in which each of these authors has remarked on the close resemblance which may be observed between the Paget cell and an aplastic melanoblast.—R. K.]

Discussion.—Dr. A. C. Roxburgh said that he had “inherited” several of these cases from Dr. Adamson. They did not behave clinically as malignant lesions; or, if they did, they could be removed before they had done any damage. At intervals he saw a patient in private who had large areas of black pigmentation on the left cheek. Twice portions of tumour in the centre had been removed and sectioned, but the surgeon had never regarded the growth as being dangerous, and it certainly had never shown any tendency to form metastases. The outlying portions were always improved by electrolysis. In other cases painting the pigmented areas with carbolic acid had been successful.

Dr. I. Muñoz hesitated to express an opinion on the section thrown on the screen because it was shown for a very short time. He, however, thought the cells which Dr. Klaver described as pagetoid looked more like typical naevus cells, in which case the condition would correspond to that of a pigmented naevus. The first section suggested the early phase in which the pigment cells contained very fine granules and appeared to be segregated before dropping into the dermis.

Pachyonychia Congenita (Jadassohn and Lewandowski).—John Franklin, M.D.

In 1906 Jadassohn and Lewandowski [1] described a condition which up till that time had not been reported, under the title of “pachyonychia congenita” or “keratosis disseminata circumscripta, tylomata and keratosiès lingue”. Their case, a girl aged 15, had a peculiar congenital condition of the nails. All the nails were hard, greatly thickened, and had to be cut with a chisel; the finger-nails were long and narrow, smooth, shiny, rather transparent, and towards the tips greyish. They were thick at their free ends and curved transversely. The toe-nails were similar and resembled an onychogryphosis. Other symptoms were pointed red papules around the nose and chin. Hyperidrosis of the nose, hands, and soles of the feet; bullae
on the soles of the feet; patches of keratotic follicular papules on the extensor surfaces of the hands and elbows; keratotic lesions on the palms and soles; and whitish patches on the tongue. The patient had a brother aged 4 similarly affected.

Since this first case a number of others [2-14] have been reported on the Continent and in the United States under the name of pachyonychia congenita, the essential feature being the curious thickened state of the nails, which is present at or begins very shortly after birth. The other chief lesions which as a rule appear soon after birth, are leucokeratosis of the tongue, symmetrical follicular keratoses affecting the extensor surfaces, keratoses of the palms and soles, and bullae on the feet. A number of other symptoms have been described, notably hyperhidrosis of palms and soles, herpetiform lesions round the mouth, dyskeratosis of the cornea with opacities [13], keratosis of the buccal mucosa as well as the tongue, and in one case involvement of the nasal mucosa and tympanic membranes [2]. The onset of these subsidiary symptoms may take place at any time after birth, sometimes many years later.

The present case:


Histology.—According to his mother he was born at the eighth month, when the nails only were affected. At the age of 3 weeks he apparently suffered from a severe stomatitis, which lasted several months. Shortly after this he had some generalized eczematous condition which lasted until about the age of 13; this eruption cleared up and was followed by the present horny condition of his skin. In 1923, at the age of 2, he was shown before this Section by Dr. Sequeira as a case of congenital onychogryphosis [15]. No note of any abnormal skin condition was made at that time. There is no history of a similar condition in any member of his family, thirty of whom are known personally to the boy and his parents.

The Registrar's Department of the London Hospital has kindly sent me the following note about him when he was an in-patient under Dr. Sequeira for five weeks at the age of 2:

"History.—Born with thickened skin in place of nails. One month: Impetigo first on face, now generalized.

On examination.—Solid tubular horny growths from all nail beds; 1 to 2 in. long; 1/2 to 1 in. diameter, projecting at an oblique angle. W.R. negative. Nails removed under alcohol, chloroform and ether mixture. Patient discharged, having contracted measles."

Present condition.—He is small, well developed, and of normal intellect.

(1) Nails: All nails of both hands and feet are markedly thickened, present a heaped-up appearance, and are a dirty yellow colour (figs. 1 and 2).

(2) Tongue: The tongue is irregularly splashed with smooth white patches which peel off from time to time leaving a shiny red base (leucokeratosis). The buccal mucosa is normal, as also are the teeth.

(3) Skin: This is generally mildly xerodermic with areas of follicular keratosis mainly on the extensor surfaces of the limbs, and on the hips and buttocks where many of the individual lesions are distinctly spiny. On the palms and soles are numerous large discrete keratotic lesions. On the feet, mainly round the toes, there is a tendency to the formation of bullae which rupture, leaving painful foul-smelling raw areas. On the face there is oily seborrhoea, with comedones and acne lesions. At the angles of the mouth herpetiform lesions which rupture and crust are always present. There are no keratotic lesions on the scalp which is seborrhoeic, and the hair is normal. There is no hyperhidrosis. There are many deeply pigmented moles on the trunk and limbs.

(4) Eyes: The eyes are normal; there is no involvement of the cornea. Wassermann reaction negative.

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\text{Fig. 1.—O. C.}

\text{Fig. 2.—O. C.}

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5 Schäfer, ibid., 1924, 148, 425.
7 Fox, Arch. Dermat. & Syph., 1928, 18, 794.
10 Dasso, ibid., 1934, 30, 218.
11 Sohrweide, ibid., 1935, 32, 370.