FOREIGN LANGUAGE ARTICLES.
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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.
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On the potential combinations of Sebocystomatosis Günther [Steatocystoma multiplex]¹ with other diseases
by S. Engel and Barbara Pinzer

With 10 illustrations

Summary: Report about the combinations of steatocystoma multiplex with other diseases that have been reported so far. Two of our own observations are described. In the first patient we found a steatocystoma multiplex probably combined with a degenerative disease of the brain.

In the second patient, we found both a steatocystoma multiplex and a keratosis palmoplantaris, a keratosis follicularis, an acne conglobata, and nail abnormalities.

Sebocystomatosis, keratosis palmoplantaris, keratosis follicularis, and nail abnormalities were found in three members of his family; in five members only a keratosis palmoplantaris was found. Our question is whether this coincidence is a matter of chance or whether this combination is a syndrome. Based on the results of histological examinations, we conclude that two sorts of cysts occur in steatocystoma multiplex: sebaceous cysts and retention cysts.

No references to nail abnormalities in the literature review on pp. 687-688. Description of case 1 on pp. 688-690; no references to nail abnormalities.

p. 690 – 692; complete translation of description of case 2.

Case 2. A 26-year-old man (D.S.) was admitted in March 1968 with the diagnosis of acne pustulosa. Apart from the usual childhood diseases, he claimed that he had never had a serious illness. However, he was treated for a "neck carbuncle" as an inpatient in 1963, and

¹ Throughout the translation of this article, the current reference, steatocystoma multiplex, German "Steatocystoma multiplex" will be used, although the German original uses "Sebocystomatosis" throughout the text. "Sebocystomatosis Günther" is rarely found in German texts at present.
since then has been in the care of a dermatologist on a regular basis. The clinical picture included comedo scars and follicular inflammations that were disseminated across the trunk and continued to the thighs. They appeared most frequently on the back, including the posterior beltline and buttocks. There was also an occurrence of giant and double comedones. Some of the follicular inflammations presented central necrolysis, which created the impression of furuncles. There were scars, some of which were hypertrophic and formed as bridge scars\(^2\) and corner scars\(^3\), which were the residue of follicular inflammatory processes that had already healed. The corner scars were primarily located in the area of the axillae. Multiple tumors, between the size of a lentil and a cherry, elastic and filled to bursting, occurred primarily in the anterior sweat canal, from the sternum to the epigastric angle. Upon opening the tumors, an oily-sebaceous mass drained out. The same cystic formations were found on the scrotum and on the scalp. In addition, hyperkeratoses on the soles (Fig. 2) were observed. Some were circumscribed and others were spread out. Occurrence was especially frequent on heels. Less pronounced keratinization on the palms (Fig. 3). The toenails and fingernails were thickened, yellowish, brittle, and no longer transparent (Fig. 4).

![Fig. 2. Diffuse hyperkeratoses on the soles in combination with steatocystoma multiplex; occurrence primarily on the heels (Patient D.S.)](image)

![Fig. 3. Palmar keratosis punctata\(^4\) in combination with steatocystoma multiplex (Patient D.S.)](image)

![Fig. 4. Nail abnormalities, impression of onychomycosis, in combination with steatocystoma multiplex (Patient D.S.)](image)

The results of the laboratory investigations were all in the normal range. Staphylococcus epidermidis was repeatedly grown from the pus of inflammatory acne infiltrates, some of which are colliquated, and cysts that were inflammatorily changed. We only succeeded in identifying staphylococcus aureus one time. In the culture of toenail shavings, Trichophyton mentagrophytes was growing.

In general, the picture of a steatocystoma multiplex in combination with acne conglobata included a palmoplantar keratosis (in part discrete) and nail abnormalities.

After detailed questioning, we learned that the father and grandfather, as well as the sister and brother of the patient apparently also presented one or another of these symptoms. We made

\(^2\) Literal translation of German "Brückennarbe".

\(^3\) Literal translation of German "Zipfelnarbe". The word Zipfel refers to a point, such as the pointy end of a stocking cap, or a corner.

\(^4\)KL: I assume that this corresponds to "punktierte Palmarkeratose" in original text.
the attempt to examine as many family members as possible (father, sister, and brother of the patient) and asked the family members that we examined detailed questions about other family members.

The 55-year-old father presented tumors that were yellowish, full and elastic, most of which were the size of a pea in groups on the forehead, root of the nose, and sternum and individually on the rest of the thorax, lower abdomen, and scrotum. Single giant comedones appeared on the trunk. The keratoses, some of which were warty and others spread out, and the nail abnormalities, that have already been described, were more pronounced than those in our patient. He reported that furuncles and abscesses had occurred from time to time.

The 23-year-old sister presented circumscribed verrucous and spread out keratoses of the soles, pronounced nail dystrophy on the toes and beginning nail dystrophy on the fingers. In both armpits and in the epigastric angle, there were whitish-yellowish cystic tumors that ranged to the size of a pea. On the skin of the back a follicular keratosis was localized that was easier to feel than to see. The patient's 24-year-old brother also presented this on the trunk, especially on the back and the buttocks (Fig. 5). Circumscribed papuloverrucous and spread out keratotic lesions were observed on the palms and soles. Pea- to small cherry-sized light cystic formations were located in the anterior and posterior sweat canal and the axillae.

The nails showed the same abnormalities as those of the other family members. In the culture of the nail material, Trichophyton mentagrophytes could be identified. It was claimed that the skin symptoms of all the family members had manifested in early childhood. The father claimed that 4 of his 8 siblings had had a palmoplantar keratosis (Fig. 6).

Excerpts from Discussion, p. 692 ff. follow.

p. 692, last paragraph:

In the second patient (D.S.), the grandfather had already presented the keratosis. In the next generation, in the father of the patient, the steatocystoma multiplex associated with the keratosis. In the father himself we find the palmoplantar keratosis, the steatocystoma multiplex, and acne conglobata, while the siblings have the symptoms of steatocystoma multiplex, palmoplantar keratosis, and a follicular keratosis (Fig. 6).

Fig. 6. Occurrence of palmoplantar keratosis, steatocystoma multiplex, and acne conglobata in the family of Patient D.S.

- Steatocystoma multiplex and keratosis and acne conglobata
• Steatocystoma multiplex and keratosis
• Keratosis
• Healthy

All four of the family members that were described also have the nail abnormalities in common. The question arises as to whether all these symptoms should be considered and evaluated separately, or if at least a part of them should be considered to be more than just a coincidence. In the first case, the patient himself shows the combination of steatocystoma multiplex with keratosis palmoplantaris diffusa that is punctuated on the hands and spread out on the feet, a follicular hyperkeratosis that is quite discrete or in relation to the extent of the overall condition is difficult to identify, acne conglobata, and nail abnormalities (probably in the sense of onychomycosis). In the siblings there is a concurrence of steatocystoma multiplex and keratosis palmoplantaris diffusa in the same form or dissemination, more or less pronounced follicular hyperkeratosis, and nail abnormalities (again in the sense of onychomycosis). Except for the follicular keratosis, the father presents the same symptoms. If the overall picture is put together as we have just done, we see that several confirmed dominant hereditary clinical pictures occur together that appear to associate other clinical pictures by coincidence.

If one chooses not to combine several of the symptoms, such as palmoplantar keratosis, follicular keratosis, and (with reservations) the nail dystrophy, into one syndrome, whereby the nail abnormalities in particular must be critically examined and the steatocystoma multiplex would still need to be excluded at this point, then it is not possible to classify them as one of the familiar syndromes for which palmoplantar keratosis is the cardinal symptom.

The only syndrome that could possibly be even remotely considered is the Pachyonychia Congenita Syndrome (JLS Type 1 according to Kumer and Loos [29]). These authors consider JLS Type 1 to include symmetrical palmoplantar keratosis and follicular keratoses of the skin of the body along with the pachyonychia congenita. However, this syndrome cannot be seriously taken into consideration, because our patients' nail abnormalities do not in any way correspond to those of pachyonychia congenita. However, in this context it is interesting that a concurrence of steatocystoma multiplex and pachyonychia congenita has already been described [30, 32].
We think that the occurrence of the nail abnormalities that is common to all 4 patients is striking especially when taking their age into consideration and even if both the clinical picture and the occasional identification of fungus point to onychomycosis. It is plausible that a genetically caused inferiority of the nail substance exists that could be considered to be a disposition for the mycosis, or that primary, perhaps congenital, nail abnormalities were in existence that the mycosis superimposed as a superinfection.

Next mention of nail abnormalities: p. 697, last paragraph
The occurrence of palmoplantar keratosis, follicular keratosis, steatocystoma multiplex, and nail abnormalities in 4 or 3 members of a family may be a justified occasion to consider whether three (palmoplantar keratosis, follicular keratosis, steatocystoma multiplex) or four (palmoplantar keratosis, follicular keratosis, steatocystoma multiplex, nail abnormalities) of the described and apparently partly regularly or irregularly dominant hereditary abnormalities can be collected together as a ternary or quaternary combination in the sense of a syndrome with palmoplantar keratosis as the cardinal symptom. By analogy with Sapuppo [26], we would like to suggest the name "Hereditary epidermal dysplasia with cyst formation".