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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.
Fff—Steatocystoma Multiplex (Multiple Sebaceous Cysts) with Familial Incidence in the First Case

Case 1. H.F. (JHH 1379451), a white female born in 1912, had recurrent cystic subcutaneous swellings since the age of 10 yr. These were predominantly on the scalp, but also on the trunk. Despite surgical removal of lesions since age 15 yr, fresh swellings continued to appear. Histology of 9 such lesions removed at Lutheran Hospital, Baltimore, in 1956 and 1957 showed them to be simple sebaceous cysts.

Apart from rheumatic fever with carditis at 6 yr of age, her general health had been good. She denied rectal bleeding but did have intermittent loose green stools, as well as bouts of right upper quadrant abdominal pain attributed to gallbladder disease. She had refused rectal or colonic investigations.

The pedigree shows a clear autosomal dominant pattern of inheritance of the sebaceous cysts, though the affection in earlier members is not well-documented (Fig. FFF-1). The original afflicted member is said to have been born in Germany. The patient's mother (Z.K., JHH 1077572) who died at age 67 after a stroke, was seen at this hospital with numerous sebaceous cysts of the scalp, 1 which underwent malignant change after being left in an ulcerated and infected condition for several years (Koonta, A. R.: Amer. J. Surg. 105:677, 1963). She also had a solitary rectal polyp removed, but had no evidence of colonic polyposis. In her, no bony tumors were detected by examination or x ray. No family members are known to have died from bowel cancer.

Physical examination (1970) showed numerous cystic swellings beneath and attached to the scalp (Fig. FFF-2), the largest about 2 cm in diameter. Many surgical scars were present. Two similar small lesions were present on the left chest. There were no lesions in the mouth (though 2 were said to have been removed during recent dental clearance) and no bony tumors were palpable.

Comment

This appears to be an instance of autosomal dominant multiple sebaceous cysts (similar to the families reported by Noojin et al: Arch. Derm. Syph. (Chic.) 57:1013, 1948 and Stephens in J. Hered. 50:299, 1959). The Gardner syndrome appears unlikely.

Peter S. Harper

Fig. FFF-1. Pedigree, Case 1.

Case 2. J.B., Jr., a Negro male born in 1947, presented in 1968 complaining of bumps which had appeared on the chest in the previous 5 mo. He was a construction worker. The bumps on the anterior chest, shoulders and upper arms had been asymptomatic except during heavy labor when he complained of a burning sensation in the lesions. He had always been healthy except for frequent upper respiratory infections and bronchitis.

No similar lesions were known in the parents or 5 sibs.

Examination (1969) showed numerous cystic nodules measuring 0.5-1.0 cm over the upper chest, shoulders and deltoid areas. The nodules were adherent to the overlying skin but showed no follicular dilatation. They were not tender and the overlying skin appeared normal. When a lesion was punctured, an oily, yellowish, odorless material was expressed.

Biopsy (69-870) showed a large keratin cyst lined by several layers of epithelial cells. In 1 area flattened sebaceous gland lobules were close to the cyst wall. The cyst itself contained eosinophilic material consistent with keratin.

James K. Davis

Fig. FFF-2. H.F. (Case 1). View of the occipital scalp which contained numerous cystic swellings. Several surgical scars were present.