Society Transactions

The Society Proceedings are edited by Dr. Morris Waisman, 806 First Federal Building, 220 Madison St, Tampa, Fla 33602. Transactions and photographs should be sent, in duplicate, directly to him.

DETROIT DERMATOLOGICAL SOCIETY

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Pachyonychia Congenita. Presented by the DEPARTMENT OF DERMATOLOGY STAFF of the Detroit General Hospital.

A 22-year-old Negro man has had deformed nails since birth. During infancy and early childhood he developed lesions of the glabrous skin and buccal mucosa. He has been able to use his hands well despite the deformed nails. The family history is negative. He has been in good health and is of average intelligence. The nails have been repeatedly traumatized, with resultant paronychia and loss of individual nails.

Examination showed all 20 nails to be similarly affected. The nails are thick, smooth, and shiny. The nail plate is of normal thickness and consistency, but is pushed upward by hard keratinous material collecting under it so that the distal two thirds of the dorsal surface of the nail forms a 30 to 40° angle with the axis of the phalanx. On the hands, knees, elbows, waist, buttocks, and feet are hyperkeratotic papules and plaques. There are small areas of leukoplakia on the tongue and warty papues of the buccal mucous membrane.

Routine laboratory examinations are within normal limits. Several avulsed nails show normal nail plate and proximal matrix. The terminal matrix produced a large amount of abnormal keratinous material. Biopsy from a lesion of the left buttock showed papillomatous and hyperkeratotic changes and biopsy of the right thigh, follicular or poral keratosis.

Discussion

HERMAN PINKUS, MD: Dr. Edward Kelley and I reported this case a number of years ago (ARCH DERM 77:724, 1958). It is a good example of the entity pachyonychia congenita which not only affects the nails, but also has other manifestations—for instance, papillomas on the hips and the disseminated clavus-like lesions on his hands and feet. Two questions come up: what is the mechanism of the nail deformity and how can one treat it? In one case in the literature the surgeon offered to amputate the distal phalanges of all the fingers, which is a little radical. If one considers the biology of nail growth, one can at least theoretically arrive at rational treatment. The nail normally grows from a proximal matrix, travels over the almost sterile nail bed, and then is more firmly attached at the end of the phalanx by the distal matrix which forms only a small amount of keratinous material, but which definitely fixes the distal portion of the nail. Now, the nail plate in this man seems fairly normal, perhaps a little thick, but very smooth. The real disturbance seems to be that he has an unusual amount of firm, hard keratin formed under the nail which lifts up the distal end and makes it curve like an inverted "U." Actually each of his fingernails resembles a miniature horse's hoof, and this I believe is the real trouble. In the horse's hoof the outer smooth portion is formed by something very similar to our proximal nail matrix, while the massive inner portion of the hoof is formed from a matrix overlying the tip of the bony phalanx and is, therefore, called the horn of the sole. I hope that

if we excise the distal matrix—both epithelium and the underlying determining connective tissue—perhaps we can improve his nails. This is what we want to try on one nail, if the patient consents.

Granular Cell Myoblastoma. Presented by J. N. GREKIN, MD, O. D. SCHWARTZ, MD, and R. F. ELTON, MD, of the Dermatology Staff of the Detroit General Hospital.

A 7-year-old white boy noted a nodule on the right little finger one year ago. On biopsy this was revealed to be a granular cell myoblastoma. Two months ago he was seen with a nodule of the right metacarpal-phalangeal joint. A nodule excised from this area three years ago was not diagnostic. Over the past year other nodules have appeared on the flexor aspect of the right forearm, the left wrist, the left thigh, and the right side of the back. These lesions have grown slowly and are asymptomatic.

The nodules are flesh-colored, nontender, freely movable, and do not urticate. Biopsy sections show in the corium several well-defined lobulated tumor masses containing cells with round or oval nuclei and large amounts of granular cytoplasm, and occasional multinucleated giant cells.

Discussion

DR. EDWARD A. KRULL: This patient presents two clinically different lesions: one, the nodule on the left thigh is fixed to the dermis, elevated, and has a whitish center, suggesting a dermatofibroma; the others are freely movable subcutaneous lesions with slight hyperpigmentation of the surface. The biopsied tumor showed granular cells, some of them separated by a small space—a space filled with either reticulin or collagen tissue and not endothelial-lined vascular channels and septa of alveolar soft-part sarcoma.

Granular cell myoblastoma, described by Abrikossoff in 1926, is an uncommon tumor of almost all parts of the body, one rarely diagnosed clinically. The tissue origin of the tumor, the malignant potential, and the incidence of multicentricity are confused and contradictory because of inaccurate reports. Clinically, these tumors are about 0.5 to 2 cm in size and located in the subcutaneous or the submucosal tissues. However, massive tumors, 20×16 cm, and pelvic tumors weighing 600 gm have also been recorded. These tumors are skin colored, slightly brown, or reddish. Their consistency is firm and they are usually movable and painless. However, some may be tender or pruritic. Occasionally they ulcerate. Almost any part of the body may be involved. One third of the tumors are on the tongue and one fifth on the skin. Other sites are the lips, larynx, breast, female external genitalia, skeletal musculature, and brain. There is probably no sex predilection. The age of onset seems to be 20 to 50 years; but there have been reports of both solitary and multiple lesions in children, as in this case. Of the 39 cases reported by Colberg and Hubay from Cleveland (*Surgery* 53:226, 1963), none were diagnosed preoperatively. The diagnosis of granular cell myoblastoma might be considered for any nodule or tumor, especially if the growth is subcutaneous or submucosal. In the breast, similarities to carcinoma evoke danger of a mastectomy for this benign growth. Granular cell myoblastoma of the tongue and other sites may be confused with squamous cell carcinoma by the unwary pathologist because of the pseudoepitheliomatous hyperplasia in some of these tumors. Some treatment-failures of so-called squamous cell carcinomas of the tongue are on this mistaken basis. There has been no report of squamous cell carcinoma originating in granular cell myoblastoma.

What percentage of granular cell myoblastomas are multiple? If we consider all the case reports (well over 550), the incidence of multiplicity is about 7%. However, if we consider only the case reports from 1950, the incidence of multiplicity is about 16% to 20%, probably a more accurate figure. It is also probably not true that multiple lesions are more likely to be malignant.

What is the origin of the tumors? Early workers, including Abrikossoff and other investigators who did tissue cultures, stated that granular cell myoblastomas were related to striated muscles. Fisher and Wechsler (*Cancer* 15:936, 1962) felt that the origin of granular cell myoblastoma was the Schwann cell, and suggested the term granular cell