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A CASE OF SCLERODACTYLIA WITH SUBCUTANEOUS CALCAREOUS CONCRETIONS.

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A study of the literature of the subject shows the existence of a group of cases characterised by the association of sclerodactylia or more wide-spread sclerodermia with multiple small subcutaneous calcareous concretions, which in some instances, as in the earliest described case,* have at first been mistaken for gouty tophaceous (uratic) deposits. Chemical examination has, however, shown the concretions to consist of carbonate and phosphate (chiefly carbonate) of calcium, and has likewise demonstrated the absence of sodium urate. The calcareous deposits can of course, as in our own case, likewise be distinguished from uratic nodules by the help of X-rays (skiagrams). The affection is of very slow evolution, and may continue for many years without the patient's general health appearing to suffer. For a long time the fingers or toes may be the only parts affected. The process of calcareous deposition apparently commences about the same time as the sclerodactylia, but probably the calcification is really secondary to the sclerosis, and is largely dependent on degenerative and obliterative changes in the small blood-vessels. Ultimately subcutaneous calcification may occur in other parts of the extremities, and the sclerodermia also may be no longer limited to the hands and feet. In some cases, as in our own case, the onset of the sclerodactylia has been long preceded by vasomotor phenomena, such as the frequent occurrence of local syncope in the fingers (white fingers, "dead fingers") or local asphyxia (cyanosed fingers).

Inflammation frequently occurs around the subcutaneous concretions and gives rise to extreme tenderness. Ulceration often follows,

* Described by a Swiss doctor, H. Weber, see *Correspondenzblatt für Schweizer Ärzte, Basel*, 1878, vol. viii, p. 623.

and then the calcareous material is discharged in the form of a chalky fluid, gritty material, or a definite calculus. The ulcers are very indolent, taking a long time to heal. No microbes have been found in the dischage, excepting those due to secondary infection. This process of ulceration, which occurs from time to time at the affected parts, is decidedly painful, so that there is no resemblance to the "painless whitlows" of so-called "Morvan's disease," and the finger-changes met with in the so-called "Morvan type" of syringomyelia. The fingers or toes become thickened, stiffened, deformed and shiny. Sensation is blunted over the diseased portions, and the normally soft pulpy cushions of the tips of the fingers or toes may gradually become almost as hard as stone. The metabolism has, as far as we know, not yet been specially investigated in order to throw light on the tendency towards calcareous deposition.

These cases are sharply distinguished from all the following kinds of subcutaneous or cutaneous calcification:

(1) Calcification in sebaceous cysts, sebaceous adenomata, and so-called "benign epitheliomata" (A. Malherbe, W. von Noorden, etc.), and in cystic adenomata connected with the sweat apparatus (H. D. Rolleston).

(2) Calcification in lipomata, fibromata, carcinomata, and other tumours.

(3) Calcification in tuberculous caseous nodules or in other subcutaneous chronic inflammatory nodules or scars.

(4) Calcified subcutaneous cysticerci.

(5) Calcification in subcutaneous veins (subcutaneous phleboliths) and subcutaneous arteries.

(6) Calcareous metastases in the skin (J. Jadassohn).

(7) Calcification of minute subcutaneous fat-lobules ("Fettgewebs-steine") in front of the tibiæ of elderly persons.*

(8) True bone-formation in subcutaneous tissue or in scars.

* O. M. Chiari ("Ueber die herdweise Verkalkung und Verknocherung des subcutanen Fettgewebes," *Zeitschrift für Heilkunde*, 1907, vol. xxviii, Supplementheft, p. 1) agrees with E. H. Bennett ("On Calcification of Adipose Tissue," *Dublin Journ. Med. Sci.*, 1878, vol. lxv, p. 19), that the small, freely movable calcareous nodules not rarely met with in front of the tibiæ in elderly persons, especially women, are due to calcification of fat-lobules, and not to calcification in veins, arteries or lymphatics. Chiari thinks that the cause of this calcification is interference with the local blood-supply caused by chronic deforming endarteritis.

We shall make no attempt here to analyse the literature on the class of case now under consideration. This has been quite recently dealt with by G. Thibierge and R. J. Weissenbach in their admirable monograph on the subject.* No similar cases have, as far as we know, been described in English or American literature. Yet it is quite possible that certain cases classed as examples of chronic Raynaud's disease with hypertrophic sclerotic reaction of the soft parts, and terminating in a condition of sclerodactylia with shortened, thickened, hard, deformed, scarred fingers, may really have been examples of the same affection, and the extruded calcareous material may have been mistaken for fragments of necrosed bone.

The patient (E. M. D—) in the present case is an unmarried woman, aged 50 years, of medium height, rather thin, active in her habits, and mentally normal. She looks rather younger than her age, and the menopause has not yet arrived. As a child she used to be subject to red hands and to chilblains in her hands and feet, that is to say, she might be said to have had a so-called "bad circulation in her extremities." When about twenty years of age she began to be often troubled by her fingers "going dead"; they used to become white ("local syncope"), not purple or black ("local asphyxia"). This occurred especially in cold weather, and she formerly often bathed them in hot water to take away the numbness. After exposure to cold the sensation of "pins and needles" was sometimes very disagreeable when her fingers were "coming to" again. Since the age of thirty-five years the skin and soft parts of her fingers have been gradually becoming harder and stiffer so as to constitute a condition of sclerodactylia. From time to time indolent, painful ulceration has occurred on the fingers, chiefly at the tips, giving rise to a gritty discharge. In April, 1911, Dr. Scholefield removed a little subcutaneous concretion from the back of the middle segment of the fourth finger of the right hand, and found it rather a difficult task because it was firmly embedded in a dense meshwork of fibrous tissue. About three years ago an area of swelling and redness appeared over the left olecranon. This ulcerated and was scraped out by Dr. Scholefield, who chemically examined the gritty material which he removed, but with negative result in regard to the presence

* G. Thibierge and R. J. Weissenbach, "Concrétions calcaires sous-cutanées et sclérodermie," *Annales de Derm. et de Syph.*, Paris, 1911, vol. ii, pp. 129–155.

of urates. Excepting for these troubles and appendicitis (for which the appendix was successfully removed ten years ago), she has on the whole enjoyed good health. She has never been out of England except for visits to the Continent. There is no history of any similar disorder in the family. Her father died at sixty-four years (of cholelithiasis?), and her mother died recently of pneumonia at over eighty years of age.

Present condition.—The ends of the fingers have the characteristic appearance of sclerodactylia, and in addition to that there are scars from which gritty material has been extruded. The terminal segments of some of the fingers appear shortened. The flesh over the flexor aspects of the terminal phalanges of the second, fourth and fifth fingers of the left hand seems particularly altered and feels quite hard as if subcutaneous calcification had taken place. Indeed, the tip of the little finger is white and looks as if some calcareous material was shortly going to be extruded. Owing to the thickening and hardening of the soft parts, movement in some of the finger-joints has become diminished. There is no true dissociation of sensation anywhere, but all sensation is somewhat blunted over the portions of the fingers where the skin has become callous.

The presence of calcification in the fingers is confirmed by examination with Roentgen rays, as it also has been by means of incision. Skiagrams of the left hand (see illustration) reveal considerable calcification in front of the terminal phalanges of the second, fourth and fifth fingers, and likewise a little at the tip of the thumb. In skiagrams of the right hand (see illustration) one can make out a little subcutaneous calcification in several fingers (this is, unfortunately, not all shown in the skiagram selected for illustration), less than in the left hand, but not so confined to the terminal segments. The distal portion of the terminal phalanx of the right middle finger has disappeared.

The patient's feet show nothing abnormal either by ordinary examination or by skiagraphy. There is apparently no sclerodermatous change anywhere except in the hands (sclerodactylia). The pinnæ of the ears show no concretions of any kind. There is no thickening of the ulnar nerves. There is slight redness and swelling over the right olecranon. This has only recently been observed, and resembles the redness and swelling over the left

FIG. 1.—Skiagram of left hand, showing subcutaneous calcareous concretions in front of terminal phalanges.

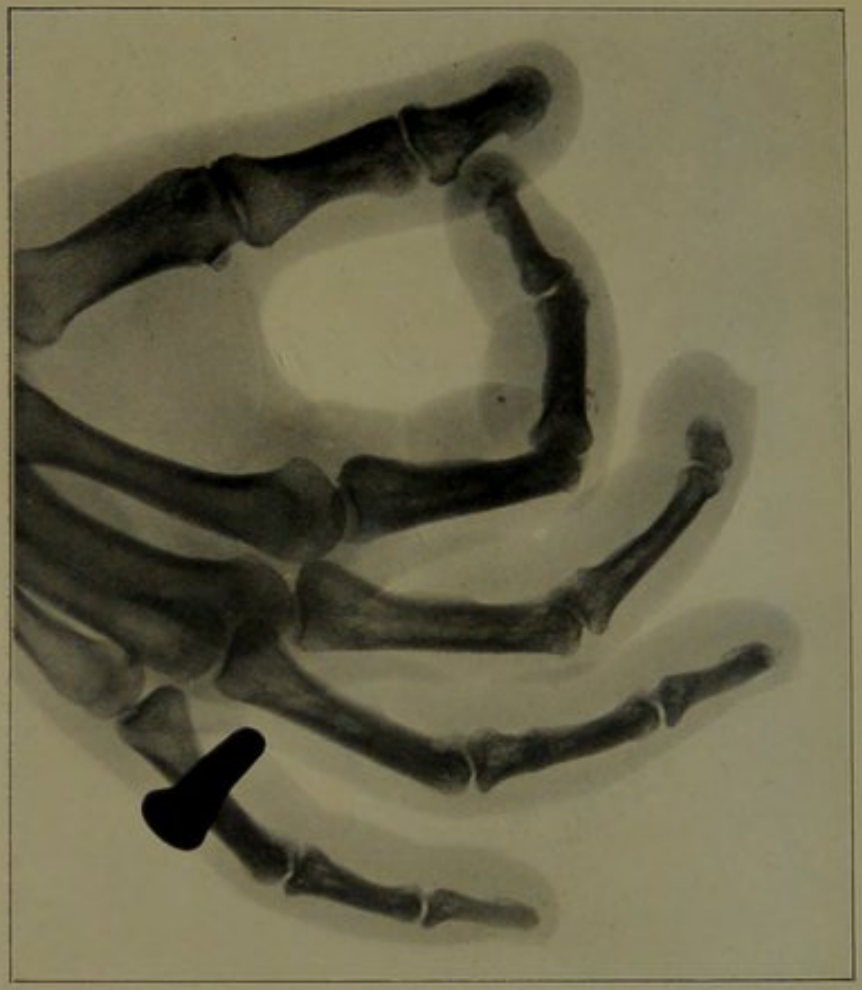

FIG. 2.—Skiagram of right hand. Note calcareous deposit at the tip of the index finger.

TO ILLUSTRATE THE ARTICLE ON SCLERODACTYLIA WITH SUBCUTANEOUS CALCAREOUS CONCRETIONS BY DR. R. E. SCHOLEFIELD AND DR. F. PARKES WEBER.

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olecranon, which, as has been already stated, occurred three years ago, and resulted in a discharge of gritty material. Skiagrams of both elbows show nothing abnormal except a minute calcareous speck over each olecranon. The knee-jerks are natural. There is no evidence of any disease of the thoracic or abdominal viscera. The urine is free from albumen and sugar. No examination of the patient's metabolism has yet been made. There is no evidence of any congenital or acquired syphilitic taint.

That the subcutaneous concretions in this case are not of the nature of gouty (tophaceous) deposits is abundantly proved by their opacity to Röntgen rays, and by the fact that Dr. Scholefield examined the gritty discharge from the left olecranon with negative result in regard to the presence of urates.

The appearance of the fingers to some extent reminds one of the stumpy thickened fingers in the so-called "Morvan type" of syringomyelia, but there is no "dissociation of sensation," and the ulceration (giving rise to the gritty discharge) has been associated with great local tenderness and has been therefore quite different to so-called "painless whitlows."

In conclusion we have to thank Dr. G. Dorner for the excellent skiagrams which he kindly made for us.