Two cases of Raynaud's disease with ocular symptoms : one case complicated by scleroderma / by George Howard Fox.

TWO CASES OF RAYNAUD'S DISEASE WITH OCULAR SYMPTOMS. ONE CASE COMPLICATED BY SCLERODERMA. ∴ ∴ ∴

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TWO CASES OF RAYNAUD'S DISEASE WITH OCULAR SYMPTOMS. ONE CASE COMPLICATED BY SCLERODERMA.

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In a recent number of the *Medical Review of Reviews* I reported three cases of gangrene presenting more or less typical phenomena of Raynaud's disease. I hesitated, however, to entitle that report Raynaud's disease, as without doubt one and possibly two of the cases were examples of peripheral endarteritis. The two cases which are the subject of this report appear to me, however, to be genuine cases of the so-called Raynaud's disease, that is, cases of symmetrical gangrene due to vasomotor disturbance, and not apparently to any organic lesions. Case No. 1 was shown by Dr. Allen before the New York Dermatological Society in November, 1905, and by Dr. Gottheil before the Academy of Medicine in April, 1907. I first had the opportunity of seeing this patient at the Vanderbilt Clinic in the service of my father, Dr. George Henry Fox. Her history is as follows:

Case 1—Feb. 10, '06; Mrs. M. J., aged 29 years, married, Russian.

Family history—Father living; mother died of appendicitis. One brother died in childhood. Three brothers and seven sisters living and healthy. There has never been any disease like the present one in her family.

Previous history—Measles, scarlatina and whooping cough in childhood. Ten years ago the patient is said to have had malaria. She remained in bed a few days, and recovered entirely in two weeks. She suffered later from a similar attack. Five years ago there was an abscess of the right ear. Paracentesis was done and was followed by a good recovery. The patient has never to her knowledge suffered from any venereal disease. With the exception of the exanthemata of childhood, she has never noticed any cutaneous eruption. The urine has never appeared abnormally dark in color.

Menstrual history—Patient first menstruated at 14. Menses occurred twice during the first year, and three or four times during

the following year. Since then her periods have been regular. They occur every 24 days, and last three to four days. Flow is moderate, pain considerable. She frequently becomes hysterical and faints just before her periods. Two years ago she had a miscarriage. Since then she has had occasional pain in the lower abdomen, and more or less continual pain in the back.

Present history—Five years ago, during the summer, the patient opened an ice cream parlor, and was in consequence compelled to constantly handle cold objects. After several months of this work she began to notice that the ring and little fingers of both hands frequently became "dead," that is, cold, numb, and white. The attacks were at first confined to the last phalanx, but later involved the others as well. Toward the end of the following winter all the fingers of both hands began to exhibit the same attacks. Since then the disease has existed more or less continuously up to the present time. The condition is always worse in winter, especially on stormy days, and better in summer. At times during the summer months she is entirely free from attacks for a few days. While the attacks are usually brought on by the application of cold water, she has found during the past year that hot water has the same effect, though to a lesser degree. The attacks are also brought on by excitement, worry or hard physical labor. They always begin in the tips of the little fingers and spread to the hand. They occur generally in the morning, less often in the afternoon, and practically never in the evening. Their duration varies from five to thirty minutes. The entire hands become reddish or bluish and swollen when the spasms of "dead fingers" have ceased. Pain during the attack is very moderate. During cold weather the hands at best feel "heavy" and more or less stiff. The spasms are confined as a rule to the fingers, the thumbs generally escaping.

Two years ago during the winter the patient began to notice on the tips of both little fingers, small scaly spots, which were painful enough to interfere with sleep. After three or four weeks the spots would become black, the pain at the same time ceasing. After several months the black tissue would fall and leave little pits. These slight depressions are sufficiently tender to interfere with fine movements of the fingers such as sewing. Last winter a similar condition was noticed in the tips of the first and second fingers. When the tips of the fingers first showed this condition, the pain was very severe and radiated to the shoulder.

The three middle toes of both feet frequently become white and cold. The great toes have never become white to the patient's knowledge, but in winter are the seat of more or less continuous pain, especially about the root of the nail.

During the last two winters the tips of the ears have become "dead" on exposure. The tip of the nose on a few occasions has become stiff and cold. At times the cheeks and chin become mottled with reddish and bluish areas, and at the same time are swollen and stiff.

On two occasions the patient noticed that the tip of the tongue became stiff for a short time.

Several times a year at intervals the patient has had attacks of dizziness and blurring of vision. At such times reading becomes impossible. These attacks last about an hour as a rule.

June 27, '06—Her condition is now worse than it has been during any previous summer, though of course it is not as bad as in winter. She feels nervous, suffers from nausea in the morning upon rising, and vomits occasionally after eating. Recently the attacks of "dead fingers" have included the thumbs.

Physical examination—Patient is well nourished, of a short, stout build. Marked internal strabismus of right eye. Pupils react to light and accommodation. Heart action is regular and there are no murmurs. Lungs apparently normal. There is more or less general abdominal tenderness, especially high in the epigastrium. The urine shows nothing abnormal.

The appearance of her face, hands and feet has varied greatly on different occasions on which I have seen her. On her first visit to the Vanderbilt Clinic (on a cold, stormy day) she presented a most classical picture of "dead fingers." The fingers were icy cold, numb and of a leaden, cadaverous appearance. The radial pulse on this occasion was also markedly reduced in volume. On subsequent occasions she has presented varying degrees of coldness and whiteness of the fingers. The spasms have invariably passed off after remaining a few minutes in a warm room. For this reason, repeated attempts to photograph the hands during a spasm of "dead fingers" have failed. Except upon her first visit the radial pulse has never shown any reduction in volume and has always been regular. There has been no increased arterial tension. There is no appreciable thickening of the arteries. The index fingers show a slight tapering of their distal portions, and the tips of all the fingers except the ring fingers present slightly thickened, depressed areas.

April 24, '07—During the past winter the thumbs have taken part in the paroxysms, being alternately whitish and bluish. The ears and nose are at times cold and white, and at others bluish. During the past year she has at times had pain in the breasts, and upon examination has found the nipples to be white and bloodless.

Finally, it may be mentioned that the patient's husband is being treated at the present time for a tubercular syphilide upon his right

elbow and forearm. The patient herself shows no evidence whatever of any former syphilitic infection. Examination of the patient's only child, a boy of nine years, fails to show any stigmata of hereditary syphilis.

A symptom of considerable importance that occurred in this as well as in my second case was sudden paroxysmal impairment of vision. I regret to say that it was not possible in either case to witness a spasm of the retinal vessels such as Raynaud and others have described. It was difficult to control my first patient, and I never succeeded in having an ophthalmoscopic examination made during one of her visual spasms. The second case had not exhibited any visual disturbances since she came under my observation, and an ophthalmoscopic examination kindly made for me by Dr. H. W. Wooton showed, as was expected, a normal fundus.

In a small number only of cases of Raynaud's disease have similar visual disturbances been described. In two of Raynaud's cases such symptoms occurred. In his first case the ophthalmoscope showed a distinct narrowing of the central artery of the retina. This did not coincide with the spasm of "local asphyxia" in the fingers, but appeared as they were returning to their normal color. Marked pulsation of the veins was also noted. In Raynaud's second case, the contraction of the retinal artery and the resulting obscuration of vision coincided with the onset of cyanosis in the fingers.

Local asphyxia and visual disturbances were combined in three cases reported by Calmette and quoted by Munro. In two cases there was narrowing of the papillary arteries, and in the third venous pulsation. All three patients had suffered from malaria. In Moriez's case the right fundus was pale, the vessels hardly visible and the papilla indistinctly limited. Narrowing of the retinal vessels was also observed in the unusual cases described by Weiss. In Bland's case, that of a maniac who would expose himself to cold by remaining out of bed, there were dimness of vision and inability to read. The ophthalmoscope showed the fundus to be unusually pale, the vessels blanched, and "almost indistinct." In the case recorded by Morgan there was considerable narrowing of the arteries of the retina, although not paroxysmal in character. In Stevenson's case there were attacks of sudden complete loss of vision, not long enough, however, to admit of ophthalmoscopic examination.

Another point of interest in my first case was the involvement of so many regions, including the fingers, toes, nose, ears, cheeks, chin, tip of the tongue, and nipples.

With regard to etiology, while cold seems to have been the exciting cause of her malady, the real cause remains unknown. There is no evidence that the patient has suffered from syphilis, even though her husband at present shows a tubercular syphilide of long standing.

Therapeutic attempts have unfortunately failed to give the patient any appreciable relief. These have included electricity, antisyphilitic treatment, nitroglycerine, etc.

My second case, whose history follows, has been for some time in my father's service at the New York Skin and Cancer Hospital.

Case 2—March 10, '06; Mrs. S., aged 50 years, single, native of Sweden; occupation, cook.

Family history—Parents died at advanced age of unknown maladies. Four sisters living and healthy. A brother died of dropsy. One sister died in childhood from fever. No disease like present one in her family.

Previous history—Patient was always delicate as a child. At seven years of age, after having eaten some alleged poisonous berries, she was attacked with severe pains in the stomach, followed by vomiting. For about twelve years following this illness she had more or less continuous gastro-intestinal disturbance. She vomited frequently after eating, and had frequent attacks of diarrhœa.

Present history—When she was about twenty-four years old both of her hands were frost-bitten, all of the fingers being affected. They were treated by friction with snow, and in a few days returned to their normal condition. For the following ten years her fingers showed nothing abnormal with the exception of frequent painful cracks on the palmar aspect. She then began to notice attacks of "dead fingers" in the last phalanges of all the fingers. She is uncertain as to the involvement of the thumbs. During the attacks which were brought on by exposure the parts involved would become cold, white and numb, and remain so until the patient got to a warm room. The attacks were of frequent occurrence.

Twenty-four years ago the patient came to this country. Three years later "blood blisters" began to appear on the tips of the fingers and some of the toes. These would rupture and scab over. Some of these lesions developed into deep-seated "sores" upon the finger ends, exposing the bone. They were extremely painful. Removal of the blackish scabs by applying a "drawing" plaster would be slowly followed by healing. The nails were frequently shed.

About ten years ago the fingers began to be stiff and the overlying skin to be drawn and tight. Three years ago the hands were exposed to the X-rays for treatment (numerous sittings). During this treatment she received a burn which confined her to bed for seven weeks and occasioned severe pain. As a result the stiffness of the

fingers became worse, especially in the right hand, which now shows marked contractures.

At the same time that the tips of the fingers presented the "sores," as described above, the fingers began to show a marked tendency to become bluish or purplish on exposure to cold. From that time to the present there have not been any such marked attacks of "dead fingers" as formerly. The terminal phalanges do not become absolutely blanched and anæsthetic as they did during the first part of her illness. They now become bluish and numb on exposure to cold.

The patient does not remember that the toes ever became blanched and "dead." The toes began to become bluish and numb on exposure to cold at the same time that the "sores" appeared upon the fingers. She also noticed at this time an eruption upon the ankles, which was moist and pruritic. This eruption recurred frequently, lasting a month or so, and disappearing under treatment.

Eight years ago there was a small sore on the tip of the last phalanx of the middle toe (right foot), which was deep and painful, and which healed after a few months.

About five years ago there was a deep sore on the tip of the last phalanx of the first toe (left foot), the diameter of a pencil and extremely painful. This healed after having lasted the entire winter. The nose and ears become cold upon slight exposure.

About five years ago the patient first noticed attacks of sudden impairment of vision. During these attacks, which lasted only a few minutes, seldom more than five minutes, all objects appeared blurred, and reading became impossible. Between the attacks vision with the aid of glasses was fairly good. The attacks continued at intervals of a fortnight or oftener for a year, and then ceased. Since then she has not had any further visual disturbances.

The patient has never noticed the urine to be particularly dark in color, or at all abnormal as far as she could judge.

Since the beginning of her illness she has lost about forty pounds in weight.

Physical examination—Patient is small and poorly nourished. Heart and lungs are normal. The radial pulse is regular and of medium size. There is no apparent thickening of the artery. There is no pulsation in the ulnar arteries and none in the posterior tibials. The moderate amount of swelling of the feet that is present might, however, easily account for the lack of pulsation in the latter vessel. The urine shows a moderate amount of bile pigment and indican. No albumen or sugar.

The left carpus forms a convexity posteriorly from side to side. The first metacarpo-phalangeal joint is partially ankylosed, and the

interphalangeal joint of the thumb completely so. The last phalanx of the thumb is clubbed and presents on its palmar aspect a superficial healing ulcer. The first finger shows a slightly stiff metacarpo-phalangeal joint. The second and third phalanges are short and are ankylosed. A portion of the nail remains. The first interphalangeal joint of the second finger is somewhat stiff, the second completely ankylosed. The last phalanx is short and rounded with a rudimentary nail. The third and fourth fingers show less ankylosis and less atrophy of the last phalanges. Nails are better preserved.

The metacarpal bones of the right hand (Figs. 1 and 2) form a slight convexity posteriorily from side to side. The thumb shows stiffness of the metacarpo-phalangeal joint and marked ankylosis of the interphalangeal joint. The terminal phalanx is atrophied and clubbed. There is ankylosis of all the metacarpo-phalangeal joints of the fingers so that the latter are held in a position of flexion. The first finger can be almost fully extended. The ankylosis increases to the little finger, which can only be extended 120 degrees. Between the first and second phalanges of the first finger a very slight motion only is possible. A small nail is preserved upon the remnant of the terminal phalanx. The middle finger shows partial ankylosis in a flexed position of first interphalangeal joint, and almost complete ankylosis of the second inflexion at 30 degrees. Nail is fairly normal. Third finger shows partial ankylosis of first and second phalanges at slight flexion. The second phalanx is conical in shape, tapering almost to a point. The third phalanx is lacking. The fourth finger shows ankylosis of all the phalanges in a flexed portion, with preservation of the nail.

The left foot presents a marked hallux valgus. The last phalanx of the first toe is decidedly clubbed, and presents on the plantar aspect a superficial scar. There is brownish pigmentation and slight œdema about both ankles. There is a moderate hallux valgus of the left foot. The last phalanx of the middle toe is clubbed.

The skin covering the thumb and fingers of both hands is smooth, glossy and mottled. It is tightly stretched and cannot be pinched up between the fingers. After slight exposure the hands remain cold to the touch for some time, and exhibit a mild degree of " dead fingers " in the last phalanges.

The sensation of pain is slightly lessened in the fingers. Sensations of heat and cold are practically normal. The constant and faradic currents are felt less acutely in the fingers than in the thumbs.

The skin covering the forearms is more or less stretched and stiff. It is smooth and there are no wrinkles or folds. The skin of the forehead and chin is smooth and somewhat " tight." The face has a more or less fixed expression. The tongue cannot be fully pro-

truded. There is general atrophy of the subcutaneous fat and of muscle. Movement in the elbows is restricted, complete extension, especially in left elbow, being impossible. There is no restriction of movement in any other joints except the fingers.

This case, in addition to exhibiting the visual phenomena above described, is interesting on account of its complication with scleroderma.

Three types of this latter affection are generally recognized. Hardy divides the disease into the (1) œdematous form; (2) scleroderma in patches and bands; and (3) scleroderma of the extremities. Thibierge speaks of generalized, progressive, and localized scleroderma. The generalized form is the œdematous variety of Hardy, where the disease begins as a more or less generalized hard œdema, this type being frequently seen in children. The localized form refers to scleroderma in patches and bands, otherwise known as morphœa. The progressive form is the one that attacks the extremities, and is frequently associated with Raynaud's disease. It is this latter form of scleroderma, often called sclerodactylia, which alone concerns us at present. The name sclerodactylia, as Thibierge points out, does not refer to a process solely involving the fingers, but to the type of scleroderma that begins in the fingers and later frequently involves the upper extremities, face, and portions of the trunk.

The relation between scleroderma and Raynaud's disease is the subject of an inaugural thesis by Favier. Both diseases, as pointed out by the latter, are found chiefly in the upper extremities, the toes being very rarely attacked. The fingers frequently assume a tapering form, the terminal phalanges atrophying, and may be reduced to one-third or less of their former size. The skin apparently adheres to the bones. The nails are partly destroyed, and the articulations become deformed. The skin assumes a violaceous aspect, and finally bullæ and superficial ulcerations make their appearance.

In his thesis, Favier quotes fourteen cases where scleroderma and Raynaud's disease were associated, and concludes that there is a close relationship between the two affections. They are both met with frequently in the female sex (11 out of 14 were women), chiefly in youth and adult age. Emotion seems to be an etiological factor in both affections, the majority of the cases having occurred in nervous and hysterical persons. The exciting causes are further almost always cold, mental emotions, or uterine disorders, acting either singly or in combination.

PLATE XXII.—To Illustrate Dr. George Howard Fox's Article.

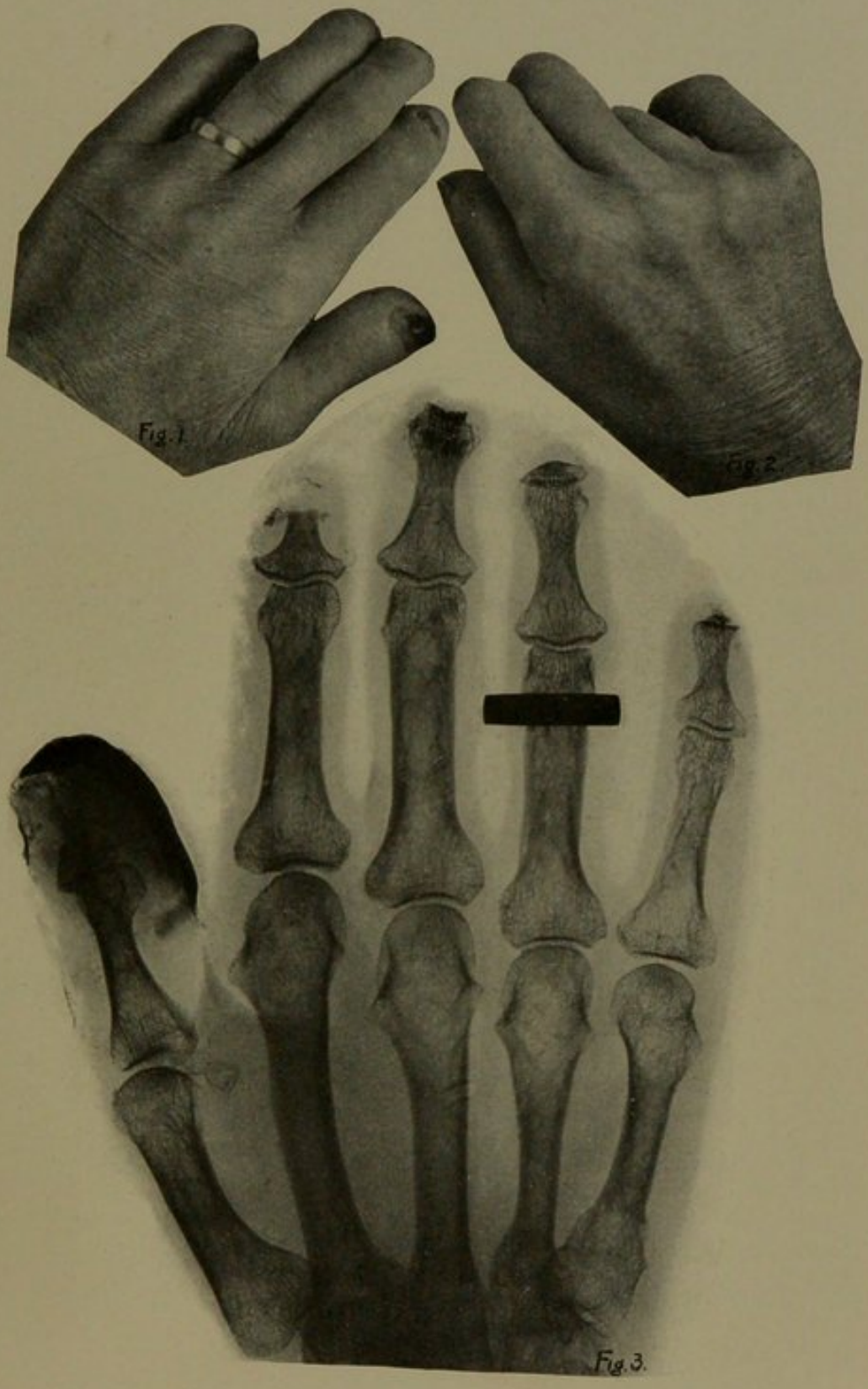

PLATE XXIII.—To Illustrate Dr. George Howard Fox's Article.

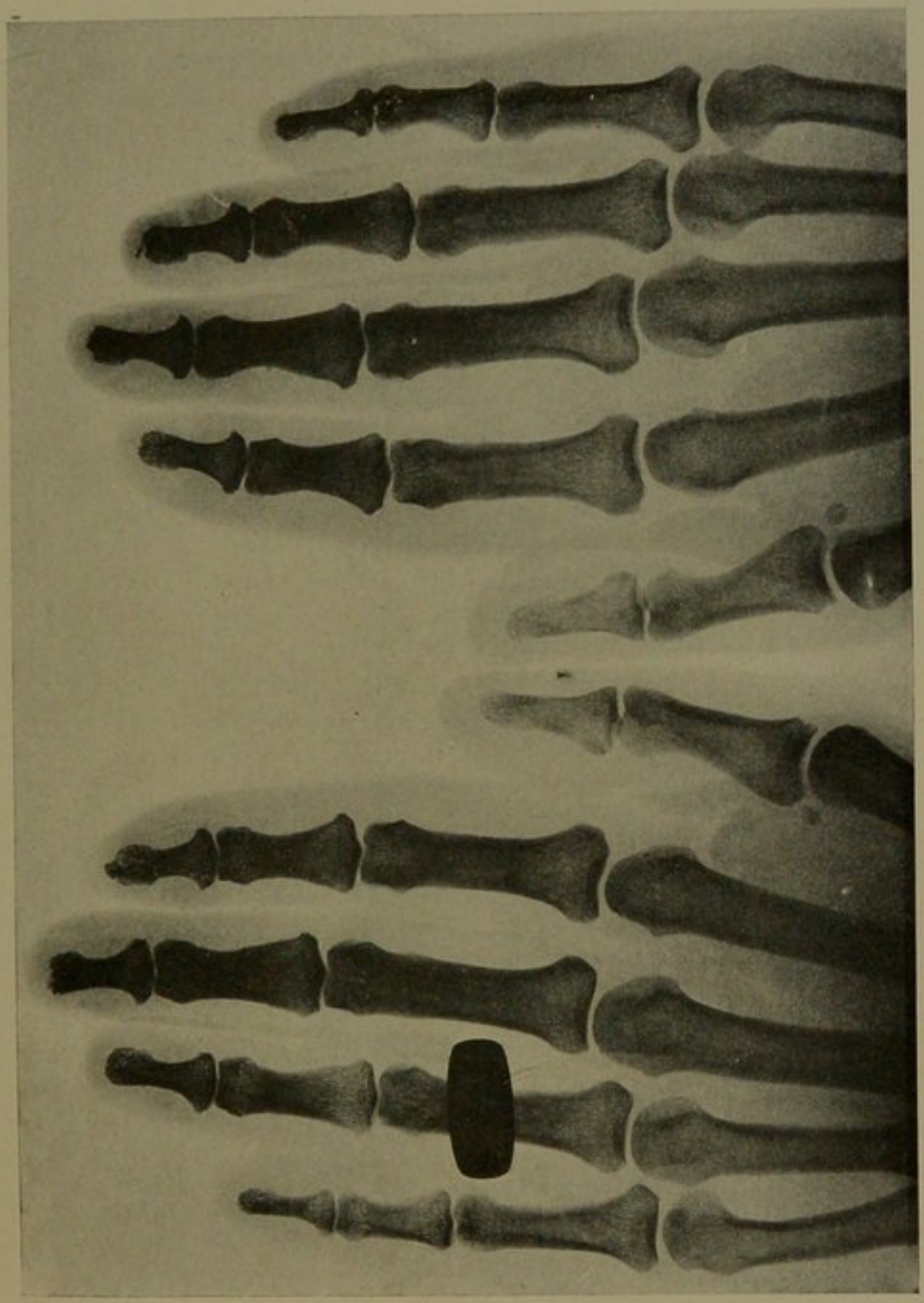

Fig. 4.

The diagnosis of the two affections from other diseases is nearly always possible, though at times difficult. Favier states, however, that it is often impossible to separate one affection from the other. "One frequently calls forth the other, and if they are met with singly it is at a slightly advanced period of the malady, when the lesions have not had time to make their appearance." In speaking of a case exhibiting symptoms of both scleroderma and Raynaud's disease, Grasset says: "One can only draw the conclusion that scleroderma and local asphyxia have close affinities. The two diseases depend upon a pathological state, if not identical, at least closely related."

In his excellent monograph on Raynaud's disease, Munro writes: "Either Raynaud's disease or scleroderma may exist for an indefinite time, and either may be complicated after a time by the supervention of the other." Munro has collected 180 cases of Raynaud's disease, and of these, 13 cases, or 7.2 per cent., showed more or less marked tendency to scleroderma. All of these cases were females. While both Raynaud's disease and scleroderma affect women by preference, "the occurrence of the latter," writes Munro, "as a complication of the former appears to be relatively still more closely restricted to females." The sclerotic changes may begin about the same time as the vaso-motor symptoms, or a number of years later.

With regard to etiology, the real underlying cause in this case, as in others of its kind, is unknown. Cold can hardly be said to have acted as an exciting cause in this case. While it is true that the patient suffered from a frost bite of the fingers as a young woman, there was a lapse of ten years between this accident and the onset of Raynaud's disease. The long gastro-intestinal illness of her childhood may have been a predisposing cause of her malady.

Attention is finally called to the bony atrophy, shown in the accompanying radiographs. In the discussion of Case 1 at the meeting of the Dermatological Society quoted above, Dr. Piffard suggested that a radiograph might show changes in the nutrition of the bones. The radiograph (Fig. 4) which was made upon this suggestion shows beginning atrophy of the tips of the last phalanges. The latter as a whole do not appear to be thinner or rarified, but the tips, instead of being smooth and rounded, are rough and irregular. The radiograph of the left hand of Case 2 (Fig. 3) shows almost complete atrophy and disappearance of the terminal phalanges. The dark shadow seen about the thumb is due to the presence of a bandage. In my article quoted at the beginning of this report, a very marked condition of bone atrophy has been depicted.

In closing, I wish to thank Dr. George M. MacKee, to whom I am indebted for the two excellent radiographs.

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