ACROSCLEROSIS

BY

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Since Maurice Raynaud first described a condition of intermittent pallor or cyanosis of the extremities, cases have often been diagnosed as Raynaud's disease where pallor, cyanosis, pain, or gangrene of the hands, feet, nose, or ears happens to be present. More often than not such a diagnosis is incorrect. It is therefore considered justifiable to present three cases of acrosclerosis to renew attention to the fact that other diseases have symptoms of a somewhat similar nature to the true Raynaud's disease. Hutchinson (1887) stressed the fact that "Raynaud's phenomenon" was a syndrome occurring in many different conditions, and, more recently, Hunt (1936) has reviewed the many diseases which may resemble but are not Raynaud's disease.

Acrosclerosis is one of those conditions frequently incorrectly diagnosed as Raynaud's disease. True Raynaud's disease consists of symptoms of an *intermittent* nature. The pallor or cyanosis of the extremities is precipitated by cold, and nutritional changes when they occur are limited to changes in the skin; massive gangrene never occurs. All those diseases which may produce the phenomenon simply as a symptom must be excluded before Raynaud's disease is diagnosed.

Aetiology of Acrosclerosis

The most obvious changes in acrosclerosis are found on the face, hands, and feet. Histologically, oedema causes swelling and separation of the collagen bundles of the cutis which eventually become hyalinized. There is cellular infiltration around the vessels, and obliterative sclerosing changes involve the intima. Dermal appendages atrophy (Lewis, 1936).

In view of these changes and the fact that the cases described below have had rheumatoid arthritis or rheumatic endocarditis, it is possible that acrosclerosis may be one of the "diseases of adaptation" (Selye, 1946), and that acrosclerosis might respond to cortisone or A.C.T.H. Diffuse true scleroderma, which is a somewhat similar disease to acrosclerosis, has already been shown to respond to cortisone (McNee, 1950). Should this prove to be the case in acrosclerosis, correct diagnosis would be essential, as no other treatment alters the course of the disease.

Clinical Picture

The majority of published cases have been those of females, symptoms beginning before middle age.

Severe pain in the fingers is almost always present and the fingers become stiff and are often blue in colour, but if the hands are raised the skin becomes white and bloodless. Cold aggravates or initiates colour changes. When the patient attempts to warm the fingers they become deep red and more painful. As the patient learns to avoid cold and as the condition advances, the skin of the fingers becomes a chronic dusky red with waxy white isolated areas at the tips of the fingers. Gradually the colour changes become permanent and are no longer intermittent. The skin becomes thicker and smoother than normal. The fingers appear swollen, giving the skin a stretched, tense, shiny appearance. It is impossible to move the skin, and it seems as if it is adherent to the underlying bones and joints. Small dark areas of superficial gangrene appear on the fingers, frequently at the side of the nail. These spots are extremely painful and either heal slowly, leaving scars, often circular, sometimes linear, and sometimes star-shaped, or infection occurs and a whitlow develops. Such a whitlow is very troublesome and chronic, and results in destruction of soft tissues, the nails, or the phalanges.

Black hard necrotic tissue often builds up below and at the edge of the nail, deforming the tip of the finger. Frequently there is chronic infection below this hardened tissue, and beads of pus may ooze out at the junction of this tissue and the more normal skin. Touching the nail or the hardened necrotic tissue causes great pain. The nail and part of the finger-tip may come away as a slough, leaving a healing, scarred, and shortened finger on which no nail regrows or at most a small beak-shaped nail appears with loose edges which are cracked and turned upwards. In the centre the nail may have ridges of differing thickness.

There is gradual atrophy and absorption of the phalanges, so that each segment of the finger becomes shorter, and sequestra may be discharged from the shortened end of the finger.

Eventually, after many years, some of the fingers become shortened to a level where sufficient blood seems still to flow through the skin and no further destruction or whitlows occur. At this stage the fingers are stiff and straight, but fixed in flexion at the metacarpo-phalangeal joints, so that the hands are of little use. The scars at the end of the shortened fingers still remain acutely painful to touch. Telangiectases may appear on the hands.

The facial appearance is typical. Telangiectases are dotted over the face, being frequently a dusky purple in colour with scattered areas of pale skin here and there, so that the general appearance is one of a high colour but mottled. In these cases the skin is dry, and fine scales may be scraped from different areas. The wrinkles are still well marked, especially around the nose and mouth. The nose becomes narrowed and pinched, with dry areas of skin at the edges of the nares. The mouth is small and the lips are greatly narrowed, so that little or no mucous membrane is visible when the mouth is closed. The hard palate is high and narrow, and if the teeth are still present the upper incisors are prominent, which exaggerates the tightness of the mouth.

In others the skin of the face becomes thick, tense, smooth, and glossy. The telangiectases are then cyanotic and the remainder of the skin is waxy. These changes are more pronounced over the forehead and around the eyes and mouth, where the skin cannot be picked up with the fingers and where the wrinkles disappear. The face assumes a mask-like appearance. The nose and mouth are likewise involved in these cases, so that all these patients resemble each other in general facial appearance.

The skin on other parts of the body may be slightly affected, more so on the feet, arms, and neck. Areas of pigmentation may appear on any part of the body, or the shade of the skin may vary in different regions, making it appear unwashed. The body hair becomes scanty, the patients become flabby, and the abdomen is obese.

A previous history of rheumatoid arthritis or rheumatic fever may be obtained, in which case cardiac disease is likely to be present.

Case 1

A married woman aged 59 was admitted to hospital on May 19, 1947, with left ventricular failure. She had acrosclerosis, which had been diagnosed elsewhere as "Raynaud's disease." At the age of 47 she had been admitted to hospital with rheumatoid arthritis, which affected all the joints of her body; treatment included gold injections, and the condition subsided. Two years later her hands began to turn blue and become cold in cold weather. The fingers became sensitive to slight injury; she often had sores on the knuckles, and whitlows developed easily. The fingers became stiff and gradually flexed at the metacarpophalangeal joints, so that she was unable to grip objects. The repeated whitlows resulted in alteration of the shape of the nails. Her facial appearance changed, and many cyanotic telangiectases developed. Recently she had become very breathless, and had frequent attacks of cardiac asthma.

On examination the face showed multiple cyanotic telangiectases. The skin was cold, thick, and waxy in appearance. The mouth was small and the lips were inverted and cyanosed. The nose was narrow and very firm. There were multiple healed scars on the hands and over the knuckles; many of the scars were star-shaped, consisting of three, four, or five lines radiating from a small depression. The terminal phalanges were shortened; all the nails were deformed and very ridged in the centre, but thin and cracked at the edges. There were a few small recent trophic ulcers surmounted with a thick scab; similar ulcers were present on both elbows, and there was one on the right shoulder. The toes were cold and the skin on the dorsum of the feet was thick and tightly adherent to underlying tissue. The abdomen was pendulous and body hair was absent. There was gross enlargement of the heart (confirmed by *x*-ray examination), and gallop rhythm was present. Electrocardiograms showed incomplete bundle-branch block of the left or common type. B.P. 170/120; W.R. negative.

After one month in hospital she was allowed home, but gradually became more and more breathless and oedematous. Multiple sores appeared on all fingers, becoming septic and very painful. She died three months after leaving hospital.

Case 2

A married woman aged 40 was admitted to hospital on November 29, 1947, with early congestive heart failure. She had acrosclerosis, which had been diagnosed elsewhere as a case of "Raynaud's disease." She gave a history of rheumatic fever when aged 16, and again at 25 years. From birth she had many small cyanosed areas on her face. From childhood her fingers often became cold and cyanosed. Her fingers were very sensitive to injury and easily became septic, causing severe pain. She had frequent whitlows, and once when the middle right finger was poulticed for several days the end separated and came away with the poultice. Over a period of years slowly healing whitlows were accompanied by shortening of the fingers. Since the age of 24 she had had no further whitlows, but the slightest knock to the fingers was very painful. She had very prominent teeth as a young girl, and her mouth was always small. During her first pregnancy her skin became very dark, the pigmentation increasing with her further two pregnancies. Axillary hair disappeared and pubic hair became scanty. Recently she had become breathless on exertion, developed oedema of the ankles, and had palpitations and angina on effort.

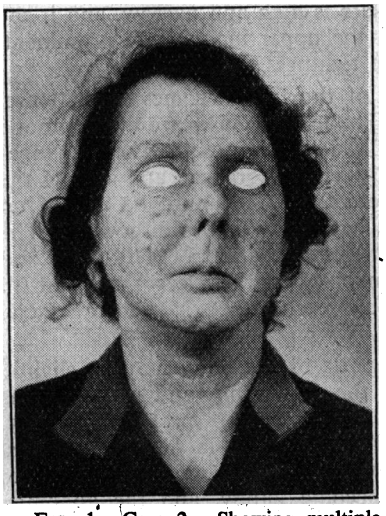

Fig. 1.—Case 2. Showing multiple telangiectases on the face, with pinched nose and narrowed tight lips.

On examination multiple telangiectases were seen on the face, the nose was pinched, the mouth small, the lips being stretched over one remaining prominent upper incisor tooth (Fig. 1). The skin on her forehead was thick and wrinkled with difficulty, and over the whole of the face the skin was waxy between the telangiectases. The fingers were stiff, the skin being very thick, hard, cold, and cyanosed. There was shortening of the terminal phalanges of both thumbs and index and middle fingers, with marked deformity of all the remaining nails. There were circular and linear scars on the finger-tips. The skin of the whole body was pigmented, especially on the back of the neck, axillae, and pubic area. Auricular fibrillation due to mitral stenosis with regurgitation was present.

Barium swallow and barium meal showed no evidence of scleroderma. Follow-up 18 months later showed no material change.

Case 3

A married woman aged 46 was admitted to hospital on October 17, 1950, with a previous diagnosis of "Raynaud's disease." When aged 34 she had developed rheumatoid arthritis affecting most of the joints of her body. A year later she was admitted to hospital, where she had treatment, including gold injections, and the arthritis subsided. When she was 38 the fingers began to feel cold and often became deep blue in colour. At other times they were a dull red. These colour changes were more frequent in cold weather. Shortly afterwards the tip of the left fifth finger became black, hard, and very painful. Two weeks later the tip of the finger became worse and finally sloughed off. Since then the other fingers had slowly developed a similar condition. Telangiectases developed on the face, the skin of which was at one time fine and soft. At the age of 43 she had a bilateral cervical sympathectomy, which resulted in the hands becoming warmer, but there was no other change in symptoms, and gradually the hands returned to their original coldness, though the rest of the arms remained warm. More recently she became breathless on exertion.

On examination there were many tiny telangiectases on the face. The skin was dry and mottled. The nose was pinched, with scales around the nares. The lips were contracted and thin, and the opened mouth was small. The fingers and thumb were shortened and deformed (Fig. 2). The skin was dull red, shiny, thick, and adherent to deep tissues. Telangiectases were present. The nails were thin and deformed, and several were raised from the pulp by

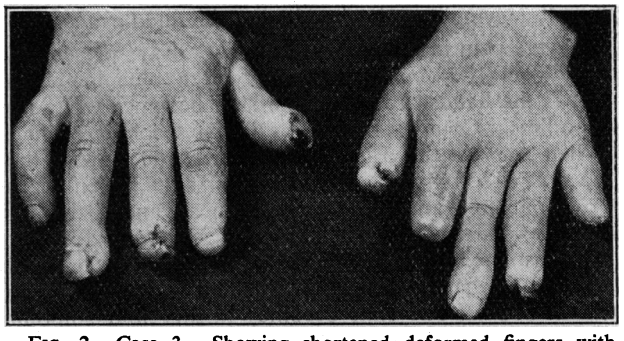

Fig. 2.—Case 3. Showing shortened deformed fingers with multiple active sores, gross deformity of the nails, and two large telangiectases at the base of the fingers on the right hand.

thick necrotic tissue from the edges of which pus constantly oozed. There was an odour of gangrene, and the nails were extremely painful to touch. Circular scars covered the ends of the fingers. The body skin was slightly pigmented, the abdomen flabby and prominent; axillary hair was absent, and pubic hair scanty. Mitral stenosis with regurgitation was present.

Discussion

Others (Harbinson, 1904; Johnson and Hedges, 1935) have reported cases in which rheumatoid arthritis preceded the onset of acrosclerosis, but few cases in the literature are described fully enough to show whether or not arthritis is a frequent forerunner of the disease. As the arthritis has involved many joints other than the hands it is unlikely that the pains and stiffness are actually the onset of acrosclerosis.

Hunt (1936) described a "rheumatic" arteritis in which sore throats, arthritis, carditis, or chorea may rapidly be followed by a typical Raynaud's phenomenon, and in severe cases "nutritional changes such as superficial sores at the finger-tips and whitlows may appear," but such cases usually improve rapidly. Hunt also thought that "the vascular symptoms and the rheumatic manifestations suggest a common aetiological factor."

Littler and Canter (1951) are of the opinion that dysfunction of the suprarenal glands is the chief aetiological factor in the pathogenesis of acrosclerosis. They quote Winfield (1904) as obtaining much improvement in a case given suprarenal extract and Schwartz (1917) as improving six cases with pituitary extract.

Two of my cases have had rheumatoid arthritis and two had rheumatic endocarditis. It is suggested that acrosclerosis is a disease of adaptation and would probably respond to cortisone treatment.

Summary

The clinical picture of acrosclerosis is reviewed and three cases are described in detail. It is suggested that it is a disease of adaptation and would respond to cortisone therapy.

References

Harbinson, G. C. R. (1904). *British Medical Journal*, **1**, 126.
Hunt, J. H. (1936). *Quart. J. Med.*, **5**, 399.
Hutchinson, J. (1887). *British Medical Journal*, **2**, 164.
Johnson, C. A., and Hedges, R. N. (1935). *Surg. Gynec. Obstet.*, **60**, 1077.
Lewis, T. (1936). *Vascular Disorders of the Limbs*. Macmillan, London.
Littler, T. R., and Canter, S. (1951). *Lancet*, **1**, 139.
McNee, J. W. (1950). *British Medical Journal*, **1**, 113.
Schwartz, P. (1917). *J. cutan. Dis.*, **35**, 256. Quoted by Littler and Canter (1951).
Selye, H. (1946). *J. clin. Endocrinol.*, **6**, 117.
Winfield, J. M. (1904). *J. cutan. Dis.*, **22**, 586. Quoted by Littler and Canter (1951).

The prefect of Paris issued a decree on June 8 introducing a compulsory medical examination of all taxi-drivers in the capital. Men up to the age of 45 are to be examined every 10 years, and every five years thereafter. The first summons to examination called upon 80 men on September 19, and about half this number attended (*Le Figaro*, September 20). The examination included visual and hearing tests, and the investigation of any propensities to blackouts or nervous troubles. Following these examinations almost all Parisian drivers went on strike for 24 hours in protest, and they have threatened to take further action if the ruling is not modified. Most of the objections are understood to come from the older drivers.

ABACTERIAL CYSTITIS

REPORT OF THREE CASES AND ISOLATION OF A SPIROCHAETE FROM ONE

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Abacterial cystitis is the name given to the syndrome in which there are symptoms and signs of cystitis but no organisms can be demonstrated in the urine by ordinary methods of examination. Characteristically, there is no constitutional upset, but the local symptoms are severe, with dysuria, frequency, and urgency of micturition, and sometimes terminal haematuria; there may or may not be a preceding or coincidental urethritis; cystoscopy reveals an intense cystitis and marked reduction of bladder capacity; and pyelography may show slight dilatation of the ureters and renal pelves. The urine is loaded with pus and sometimes red blood cells, but no organisms can be seen in the stained centrifuged deposit, and no growth is obtained on the usual culture media, either aerobically or anaerobically. Tuberculosis is often suspected, but tubercle bacilli are never found even after guinea-pig inoculation.

Local treatment of the bladder, and the use of urinary antiseptics, sulphonamides, and penicillin have little or no effect. The attack may clear up spontaneously, but in many cases the cystitis persists for months and even for years. After the administration of neoarsphenamine there is always a dramatic response and rapid cure, irrespective of the duration of symptoms. The pyuria disappears, and the bladder capacity, the bladder mucous membrane, the ureters, and the renal pelves all return to normal. Recent reports indicate that arsenoxide ("mapharside") and quinquevalent arsenicals are as effective and that streptomycin and "aureomycin" may also be of value.

It has been suggested by several observers that the causative organism may be a spirochaete. Coutts and Vargas-Zalazar (1946) stated that they found "spirilla of diverse morphology" on dark-ground examination of the urine in five cases and Fieldsend (1947) found "numerous spirillum-like bodies" in one case. These observers did not identify the organism further. It is known that spirochaetes are not uncommonly present even in the healthy genito-urinary tract, and that "spirillum-like bodies" may be seen in degenerating pus. It is difficult, therefore, to determine the significance of such findings in the urines of cases of abacterial cystitis unless the spirochaetes are more exactly defined.

Spirochaetes were seen on dark-ground examination of the centrifuged deposit of urine in three cases of abacterial cystitis recently investigated in the department of venereal diseases at Leeds, and from the urine of one of these cases a spirochaete has been cultured and an attempt made to identify it. Abacterial cystitis is shortly to be the subject of a fuller review by one of us (G. O. H.), and further studies of the spirochaete recently isolated are being undertaken, but a prelimi-