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ERYTHROCYANOSIS

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The condition of erythrocyanosis with which this paper is concerned figures in Continental literature under the title of "erythrocyanosis crurum puellarum," a title made still more cumbersome by the addition of the word "frigidum" in some writings. French surgeons and dermatologists describe it as "erythrocyanosis sus-malléolaire." The condition is, as these names imply, one of reddish-blue discoloration of the skin seen in the lower half of the leg in girls and young women, and especially evident in the region above the malleoli. In this respect it differs from acrocyanosis, "acro-asphyxie," which affects the most peripheral parts of the circulation—the digits, the ears, and the nose. Erythrocyanosis may affect the skin over the triceps in the arm, but seldom leads to any serious consequences in this situation. Examples of the disease in the male are very rare.

General Features

In its minor degrees the lesion in the legs is very common and trivial, but it may develop in some cases into a serious and even crippling disability. An advanced case shows the following features. The lower half of the leg, particularly on its posterior surface, is of a purplish hue, which may be uniform or marked by scattered plaques of a deeper colour. The colour is blanched by pressure, but when the pressure is released the area fills up quickly with dark blood from all sides in a way which Krogh has graphically likened to a subcutaneous lake of ink. The discoloured area is cold to the touch. The circumference of the limb is much increased by a firm, elastic infiltration of the subcutaneous tissue. This may reach very considerable dimensions, and may take the form of a thick and prominent cuff of tissue most

evident just above the malleoli and shading off higher up the leg. It produces a clumsy and evident deformity. The overlying skin is characteristic; it is stippled with small reddish points, in the centre of which is a minute keratinized focus. These are the hair follicles, enlarged and pigmented; the hair is usually absent, the "keratose pileaire" of French authors.

From time to time fairly large and very distinct indurated nodules appear in the subcutaneous tissue. When fully developed they are circular plaques 1 to 2 cm. in diameter, and are painful and very tender. On inspection the overlying skin appears a little tense and reddened, and on palpation a definite discrete induration is found, which spreads deeply into the subcutaneous tissue. Several patients have told us that they can forecast their appearance by a severe itching long before the nodules are apparent. These nodules undergo a slow resolution which at the best occupies several weeks. They often leave evidence of subcutaneous scarring, and in one of our patients, a middle-aged woman who had suffered for many years, there was a large area of thin, fibrous skin in the lower half of the posterior aspect of each leg. She stated confidently that at no time had there been any ulceration. On the other hand, in a number of cases a nodule here and there enlarges, becomes acutely inflamed, and finally bursts, leaving a small circular ulcer in the base of which is a tough, adherent slough. These ulcers are very slow to heal and often last until the beginning of summer. In the more severe cases sensations of weight and coldness in the leg, intolerable itching, and burning pain, together with the appearance of ulcers, may render the patient quite unfit for any work.

The thin, spare leg appears immune to this disease. Persons suffering from the condition are of a stout and florid build with large limbs, in which the sturdy appearance is misleading and due to excessive fat. Their general appearance is that of rude health, and Mach,¹ who gives a very good description of the lesion, states that they excite no pity because they look so healthy.

Aetiology

A study of the very extensive literature of erythrocyanosis shows that authors have been concerned mainly with two different views in aetiology. According to some

the lesion is tuberculous, while others postulate some endocrine dysfunction. Sections of the nodules removed from cases of erythrocyanosis show giant cells which are not of a true tuberculous type, but which may easily be reported as such unless the observer has had special experience in this field. Some workers, realizing that proof of the presence of the tubercle bacillus itself is not forthcoming, assume a tuberculous toxin as the cause. Such reasoning is not capable of direct proof, and like "la tuberculose inflammatoire" of Poncet² must be regarded as purely speculative.

The school of endocrine dysfunction incriminates variously the thyroid, parathyroid, ovarian, and pituitary glands. The build of some of the patients suggests some degree of pituitary dysfunction, and this was notably so in two of our cases. These were submitted, however, to every known test of the pituitary function, the one by Professor Stopford and the other by Mr. G. F. Rowbotham, with entirely normal findings in each case.

We believe that a clue to the aetiology of this disorder may be obtained by a study of the vascular lesions in severe anterior poliomyelitis. These lesions are clinically identical with those in erythrocyanosis. The long-standing case of severe anterior poliomyelitis presents the same purplish discoloration in the same district with the same subcutaneous nodules running an exactly parallel course. In a former paper³ evidence was adduced that the condition in anterior poliomyelitis was due to a deficient blood supply, more especially a deficient venous return, in a loose, unsupported fatty area far out in the peripheral circulation and exposed to cold. If, as we suggest, these lesions in severe anterior poliomyelitis are the same as those in erythrocyanosis, there can be no need to invoke either tuberculous or endocrine troubles as agents. It is clear that children of every conceivable diathesis are liable to suffer from anterior poliomyelitis, and if they are so unfortunate as to lose almost all muscular power in a leg they will eventually display vascular changes.

The factors which are constant in the two diseases erythrocyanosis and anterior poliomyelitis are defective venous return, excessive fat, and the action of cold, and it is in these purely local and physical factors that the explanation is to be sought. The relation to fat is very striking. Erythrocyanosis is found in women in the fat of the leg, and to a lesser extent in the fat around and above the knee, in the buttock, and occasionally in fat breasts. In the very few cases in which the lesion has been described in the male it has been seen in the buttock. The part played by exposure to cold is evident when one considers the very scanty clothing of the lower extremities which is the fashion to-day in women.

The views we have put forward agree closely with those of Klingmüller,⁴ who regards the condition as a perniosis, and states that it is a purely vascular disturbance. We would venture from our experience to suggest that the same pathology may apply also to Bazin's disease.

The clinical features of the cases suggest that in consequence of deficient venous return, assisted by the constricting action of cold, a portion of the already subnormal tissue suffers an insult which may be due to starvation or to the escape of some noxious agent, the product of retarded circulation. The reaction of the tissues causes the nodule, which ultimately either gradually subsides with terminal fibrosis or, being unable to overcome the trouble, ends in a slough, which is finally cast off by ulceration. There is a striking resemblance on both clinical and histological grounds between this nodular lesion and that of fat necrosis. One of our patients, a very stout middle-aged woman, had a tumour removed from her left breast four years ago. This proved on section to be fat necrosis. She now has an exactly

similar patch of fibrous induration in the fat of the right buttock and on the posterior aspect of the right calf.

Treatment and Results

At the beginning of this article we stated that the majority of these cases are trivial, and certainly no one would contemplate a major surgical operation for their treatment. We have, however, encountered ten cases in all in which the pain, disability, and total incapacity for work justified operative treatment. These were treated by bilateral lumbar cord ganglionectomy, and have up to the present given, without exception, most satisfactory results. The longest period which has elapsed since operation is four years and the shortest six months. The patients remain well without discoloration or other trouble, and are all fully restored to normal activity. It is to be noted that the clumsy and disfiguring cuff which forms above the malleolus is not entirely removed, but diminishes gradually until it is no more than a quarter of its former size.

Summary of Ten Cases

Case 1.—Female, aged 33. Fifteen years' history of trouble from pain, aching on exertion, and extreme coldness. Much supra-malleolar thickening. Unable to take exercise and live a normal life. Since operation on April 3rd, 1932, patient has remained entirely free from former trouble.

Case 2.—Female, aged 16. Severe chilblains and blue, swollen legs for four years. Unable to follow her work. Operation performed on April 16th, 1932. Patient remains very well and in regular employment.

Case 3.—Female, aged 17. Three years' history of cold, cyanosed legs, with severe chilblains; many scars of ulcers present. Since operation on June 16th, 1932, patient has remained entirely free from trouble.

Case 4.—Female, aged 18. Both legs deeply cyanotic and swollen for six years; could not walk far; chilblains, but no ulceration. Operation performed on November 12th, 1932. Patient remained well through two winters, but her present whereabouts are not known.

Case 5.—Female, aged 22. Ten years' history of almost total incapacity during the winter; chilblains, much pain and aching, huge supramalleolar swelling with great disability. Since operation on December 22nd, 1932, patient has been perfectly well; legs warm and of normal colour; some thickening remains.

Case 6.—Female, aged 24. Chilblains "all her life," which were present throughout the year. Considerable swelling about the ankles and a large persistent ulcer above the left medial malleolus. Operation performed on November 17th, 1932. The patient is very well; swelling much reduced; ulcer remains healed and colour normal.

Case 7.—Female, aged 17. Chilblains from childhood, lasting through the summer. Both legs blue, with many scars of old ulcers. Operation performed on February 25th, 1933. Both legs now normal; no swelling, and no return of chilblains or ulcers.

Case 8.—Female, aged 33. Fourteen years' history of cold, blue legs, with chilblains and many ulcers. Operation performed on June 17th, 1933. The patient is now very well. Since the operation she has not had a chilblain or an ulcer, and her legs are normal in shape and colour.

Case 9.—Female, aged 20. Six years' history of pain, itching, and ulceration of the legs, almost as bad in summer as in winter. Since operation on April 23rd, 1934, she has remained very well; both legs are shapely and of good colour, and she never has a chilblain or ulcer.

Case 10.—Female, aged 26. Three years' history of swelling above the ankles; ulcers formed in the winter and she was unable to work. Immediate result of operation, which was performed on September 21st, 1935, was satisfactory, and she is working through a severe winter without any trouble.

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