Widespread and Sudden Occlusion of the Small Arteries of the Hands and Feet

By R. P. Jepson, F.R.C.S.

This paper analyzes the syndrome of symmetrical digital gangrene that results from various pathologic processes. If a group of such patients is closely studied, it is apparent that among those who have suffered a sudden and widespread occlusion of the small arteries of the hands and feet some may be grouped apart by reason of the single nature of the attack and the good prognosis. Eleven such examples seen by the author are described and their differential diagnosis, causation, and treatment discussed.

Following Raynaud's thesis in 1862 many case histories of minor and major limb gangrene have been selectively described as "symmetrical digital gangrene" because of the bilateral nature of the lesions. This term is not entirely satisfactory. It is apparent that the separation of "symmetrical digital gangrene" from other forms of limb gangrene is, in many ways, artificial and that it is a clinical presentation common to many different disease processes (table 1). In addition, in many cases, sudden occlusion of the small peripheral arteries may cause profound ischemia with cyanosis rather than tissue necrosis and the term "symmetrical digital gangrene" cannot be strictly applied to this less severe group. Among patients who have suffered a widespread and sudden occlusion of the small arteries of the hands and feet, with and without gangrene, some may be grouped apart by reason of the good prognosis that the disease carries and the absence of any etiologic factor. In this paper 11 such examples are described of digital gangrene or profound cyanosis of sudden onset occurring in 2 or more limbs; many of these have been reviewed for periods up to 15 years after the acute illness, and something of the natural history of the condition is described. Some of the more severely affected patients resemble those described by Raynaud under the title of "symmetrical gangrene of benign form": Lewis and Pickering added further case histories of "bilateral gangrene of digits in the young and with infection" and comprehensively reviewed the subject to that date.

Clinical Data

A history of normal health for years prior to the occlusive episodes was common to all the patients and more especially none had noticed "white fingers" or hemoglobinuria following exposure to cold. Physical examination failed to reveal any evidence of systemic disease although 4 of the patients had an associated pyrexia; Wassermann reactions and tests for autohemagglutination were negative. The reactive hyperemia test and arteriography were used to define the pattern of occlusion in the hand and finger arteries.

Group A. Arterial Thromboses Associated with Pyrexial Illness (4 patients)

Case 1. Female, aged 2 years. This child had been admitted to the hospital 2 months prior to the vascular episode with a persistent pyrexia (100-101 F) and raised erythrocyte sedimentation rate for which no reason could be found. The onset of the vascular lesions was heralded by bluish mottled patches over the outer side of the left foot. Within 24 hours both feet were cold, blue, and edematous and the ankle pulses were not palpable, although the popliteal pulses could be easily elicited. The right and, to a lesser degree, the left hand were similarly cold, cyanosed, and swollen. The right radial pulse could not be detected, although the brachial blood pressure in that arm was 120/70 mm. Hg. Within the next few days the fifth toe on the left foot became gangrenous and was finally amputated. A small necrotic area separated from the top of the right index finger. A tibial muscle biopsy was histologically normal, and arteriography failed to demonstrate any occlusion in the major vessels proximal to the ankles. Over the next few days all the major pulses returned to their normal volume and the limbs recovered normal warmth and color. When seen 5 years later the child was in

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excellent health, energetic, and agile. A reactive hyperemia test still showed a delayed return of color to all the digits. She suffered from blue and cold hands in the winter.

Case 2. Female, aged 2 years. For 10 days prior to admission to the hospital the child was noted to be unwell and was thought by her parents to have a “cold.” On the fifth day of this illness the feet had become cold, blue, and swollen, and blood-filled blisters had appeared over the dorsum of the distal part of the foot.

On admission to the hospital the child was found to be well nourished, pyrexial (101-102 F.), with feet and hands cold, blue, and edematous. Neither the radial nor ankle pulses were palpable, and oscillometer readings above the ankle were 0. The brachial blood pressure was 130/75 mm. Hg. Within 24 hours the radial pulses returned and during the following 3 days the feet showed signs of a reactive hyperemia, becoming pink and hot. A small area of skin became necrotic on the dorsum of the right foot. When seen 3 years later the ankle pulses had not returned, the child at that time being healthy and growing normally.

Case 3. Female, aged 54 years. This previously healthy housewife developed a mild “pyrexia” while

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<td>Atherosclerosis, (\text{\textsuperscript{7, 7}}) embolism, dissecting aneurysm, thromboangiitis obliterans, rheumatic arteritis.(\text{\textsuperscript{8}})</td>
<td>Pneumonia, (\text{\textsuperscript{5}}) meningococcal meningitis, (\text{\textsuperscript{10}}) cholera, (\text{\textsuperscript{11}}) diphtheria, (\text{\textsuperscript{4}}) tuberculosis, (\text{\textsuperscript{13}}) syphilis, (\text{\textsuperscript{6}}) fever of unknown origin. (\text{\textsuperscript{2}})</td>
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<td>Frost-bite.</td>
<td>Ergot, (\text{\textsuperscript{5}}) carbon monoxide poisoning, (\text{\textsuperscript{11}}) cachexia. (\text{\textsuperscript{1}})</td>
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Table 1.—Some of the Diseases That Have Been Described in Conjunction with Symmetrical Digital Gangrene

Fig. 1. Case 3. Arteriogram of left hand. Narrowing or occlusion of the digital arteries can be seen. The radial artery is slightly tortuous and it is probable that the ulnar artery is occluded.
on a holiday. On the third day of this illness all the fingers of the hands became cyanosed and painful within a matter of a few hours. When seen 48 hours later the fingers were blue and cold, and in the left little, middle, and index, and the right middle and ring fingers, the cyanosis spread proximally to the metacarpophalangeal joints. The reactive hyperemia test and arteriography demonstrated blocks in the arteries of all the digits together with normal patency of the slightly tortuous radial arteries. The left ulnar artery failed to fill and was probably occluded (fig. 1). Gangrene developed in the left middle finger, the distal part of which was amputated. Histology of the proximal digital vessels was described as showing a "bland" thrombosis with a slight degree of inflammatory reaction. A bilateral cervicothoracic ganglionection was performed, and the remaining fingers healed surprisingly well with only minimal skin loss over their pulps (fig. 2). At follow-up examination 7 years later the patient...
complained of moderate cyanosis and occasional blanching of the fingers in cold weather but was otherwise well.

Case 4. Female, aged 67 years. This housewife developed cold blue hands and a pyrexia which lasted for a few days 6 months previous to examination. On examination, her fingers and thumbs were persistently cold and blue, the tip of the right middle finger being ulcerated. These were demonstrated by reactive hyperemia to be due to digital artery occlusion with a normal major circulation.

Group B. Acute Arterial Occlusions without Premonitory Symptoms (7 patients)

Case 5. Male, aged 23 years. This man had, up to his illness, worked as a farm laborer and as an Air Force private, occupations which had exposed his hands to all weathers without untoward effects. The illness developed 3 days previous to examination and, when first seen, both ears were blue and cold, the outer rings of each being black and gangrenous. These areas finally separated and the ears healed satisfactorily. In the hands all fingertips were dry, blackened, and anesthetic, with some patchy cyanosis proximally along the ulnar border of the arm and in the right thumb. All the toes on both feet showed gangrene at the tips and cyanosis. The peripheral major arteries in the 4 extremities were palpable and of normal volume.

A bilateral cervicothoracic and lumbar ganglionectomy were performed, following which the necrotic ends of the digits separated and healed within a few weeks. This patient returned to work as a farm laborer and has been seen at regular intervals for 10 years. During this period he has remained in perfect health except for gustatory sweating on the left side of the face and abnormal dryness of the hands, which may be attributed to the cervicothoracic ganglionectomy. He is free from Raynaud's phenomenon.

Case 6. Female, aged 54 years. This patient's history has unusual features in that premonitory episodes of localized gangrene preceded the bilateral attack (Lewis and Pickering). In brief, the right forefoot became cyanosed in January, with loss by dry gangrene of the right little toe; in July of the same year the left big toe became cyanosed and gangrenous and was followed at intervals of a month by dry gangrene of the left second and third and the right fourth and fifth toes. In February of the following year all the digits of both hands became cyanosed and cold overnight; dry gangrene developed at the tip of all these, with separation and eventual healing. Bilateral cervicothoracic and lumbar ganglionectomies were performed. When reviewed 14 years later this patient (now aged 69) has remained perfectly well except that her hands are "cold" in the winter. Pulses remained normal at the wrist and ankle throughout.

Case 7. Female, aged 48 years. This housewife awakened one morning to find the little, ring, and middle fingers of both hands blue and painful; the index fingers were involved to a minimal degree but the thumbs were spared. Areas of patchy cyanosis up to several centimeters in diameter were apparent in both feet, particularly over the instep and toes. No other abnormal signs were detected, and all the major pulses were palpable. Following bilateral cervicothoracic ganglionectomy the fingers recovered their color and warmth, although a certain amount of subcutaneous wasting occurred in the pulps. After 5 years the patient remained well and free from further attacks.

Case 8. Female, aged 50 years. Her history is similar to case 7. All digits of 1 hand became cyanosed overnight, but recovered warmth and color on conservative therapy over the succeeding days with minimal ulceration of pulps. Her course was reviewed for 5 years, during which time she remained well.

Case 9. Female, aged 30 years. One week previous to examination the left index, middle, and little, and right middle, ring, and little fingers had become painful and cyanosed over their distal halves. She was otherwise free from any associated disease. The reactive hyperemia test demonstrated a gross delay (less than 20 sec.) in the time taken for the flush to reach the tips of all the digits. Over the next few weeks the fingers returned to a reasonable warmth and color without skin loss.

Case 10. Male, aged 7 years. In December this boy awakened one morning with all the fingers of both hands deeply cyanosed: to a lesser degree all his toes were likewise affected. There was no history of exposure to cold.

An arteriogram confirmed the abnormal reactive hyperemia test and demonstrated extensive thrombosis in the digital arteries, especially to the index fingers (fig. 3). The proximal arteries in the palm and arm were normal and patent. Following bilateral cervicothoracic ganglionectomy the fingers recovered their warmth and color. When reviewed 5 years later, the boy had grown and developed normally, and except for a moderate cyanosis of the fingers in very cold weather he experiences no disability.

Case 11. Female, aged 16 years. This girl developed a sudden and intense cyanosis and anesthesia of all the toes, which eventually resulted in the loss by dry gangrene of the lateral 3 digits in both feet. The hands were unaffected and both the posterior tibial and dorsalis pedis pulses were normal. Unfortunately, it has not been possible to follow the subsequent progress of this girl.
OCCLUSION OF SMALL ARTERIES OF HANDS AND FEET

Fig. 3. Case 10. Arteriogram showing patency of radial and ulnar arteries but with multiple occlusions in the digital arteries.

DISCUSSION

Gangrene of the extremities that presents symmetrically is by no means uncommon; Morgan collected 93 case reports from the literature and since that time many more have been added. The majority of these have been clearly associated with some other disease processes (table 1), and can usually be excluded by clinical examination and by routine laboratory tests. When this is done, there remains a group of patients in whom an acute single illness is followed by a remarkable degree of recovery and it is suggested that, whereas the severe forms of this "benign" malady may reasonably be collected under the heading of "symmetrical digital gangrene," many, and possibly the majority, of the cases do not progress to gangrene. The diagnosis of digital artery occlusion as described in the present paper can only be irrevocably established on follow-up of the patients over a period of many years; it seems at present justifiable to group together such cases, whether they occur in infants or adults, with or without pyrexia, with gangrene or simply digital cyanosis.

All evidence from these patients points to the final lesion being a thrombotic occlusion of the digital or similar-sized arteries. The fingers clinically involved by cyanosis or gangrene consistently showed prolonged delay in the reactive hyperemia and arteriography confirmed the presence of luminal occlusion. The arterial circulation as estimated by the blood pressure and palpable pulses at the wrist and ankle was adequate, with the exception of the 2 young girls (cases 1 and 2). In these the major pulses disappeared, later to return in 1 of them, suggesting that a prolonged arterial constriction was present early in the course of the disease. It is possible that the final occlusive lesions are secondary to a prolonged digital and more proximal arterial spasm, although there is no direct evidence to support this view. Histologic information is limited by reason of the minor forms of gangrene that were incurred; where it was obtained (case 3), the arterial wall showed only a mild inflammatory reaction to the luminal clot.

Whatever its etiology, the natural history of the disease was similar in all 11 patients. It presented as a single nonrecurrent episode (with the exception of case 6) that reached its maximum clinical severity early in its course.
The extremities at first sight appeared to be affected by such a profound ischemia that gangrene would certainly result. Experience proved otherwise; the collateral circulation, especially in the young, was rapidly established with recovery of deeply cyanosed and even anesthetic digital skin. Gangrene where it occurred was limited in extent and “dry.”

In such cases where the digital lesions are not associated with any systemic disease, the patient may be assured that limb function will be little impaired and that the episode is unlikely to be repeated although Raynaud’s phenomenon may appear in the involved fingers.

SUMMARY

The case histories are presented of 11 patients in whom multiple arterial occlusions occurred, with or without an associated pyrexia, in the digits and ears. No evidence for an associated disease could be found in these patients. The organic occlusion of the digital arteries produced marked cyanosis of the digits, which in some instances proceeded to limited gangrene. It is suggested that similar cases of severer form have been previously described under the title of “symmetrical digital gangrene.” The disease has not recurred in any of these patients who have remained well for periods up to 15 years.

ACKNOWLEDGMENT

My thanks are due to Professor A. M. Boyd, by whose courtesy many of these cases were seen.

SUMMARIO IN INTERLINGUA

Es presentate le historias de 11 patientes in qui occurreva multiple occlusiones arterial, sin o con associate pyrexia, in le digitos e in le aures. Nulle signo de morbo associate esseva detegible in iste patientes. Le occlusion organie del arterias digital produceva marcate cyanosis del digitos, resultante, in certe casos, in gangrena limitate. Es postulate le possibilitate que simile casos de forma plus sever esseva previemente describite sub le termino de “symmetric gangrena digital.” Le morbo non ha recurrite in le patientes del presente serie. Illes ha remanite libere de illo durante periodos de usque a 15 annos.

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