Phlegmasia Cerulea Dolens

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Gangrene and cyanosis of an extremity, due to thrombophlebitis with massive venous occlusions, is an uncommon condition and is often confused with primary arterial occlusive disease. With massive venous occlusion and the associated arterial venospasm the blood flow may be so impeded to an extremity that cyanosis and eventually gangrene may occur. The recognition of this is important, since this gangrene is usually superficial or limited to the digits. Conservative treatment of this type of gangrene is indicated in contrast to the more radical treatment advocated for gangrene due to primary arterial insufficiency. This disease has been called "phlegmasia cerulea dolens," "blue phlebitis of Gregoire," "acute massive venous occlusion of the extremities," "pseudoembolic phlebitis," and "gangrene of venous origin." That gangrene of an extremity can be of venous origin was first recognized by Fabricius Hidannus in 1593. Excellent review articles on phlegmasia cerulea dolens have been published by Haimovici and DeBakey, making a detailed review of the literature unnecessary. Of the 102 cases in the literature in which information is available, 69 occurred in the left lower extremity, 27 in the right lower extremity, four in the left upper extremity, and two in the right upper extremity. The high incidence in the left lower extremity is probably because the right common iliac artery crosses over the left common iliac vein shortly after the bifurcation of the abdominal aorta. Spontaneous involvement of the upper extremity is rare, and it has been reported in only five previous cases. It is the purpose of this paper to report two cases, one with involvement of the upper extremity.

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Case Reports

Case 1

A 30-year-old white man was first admitted to The Graduate Hospital on July 6, 1959. He indulged in excessive alcoholic intake for several years, with a very poor dietary intake, and he had been in chronically poor health. At 5:00 a.m. he was awakened from his sleep with pain in the left calf and thigh. The pain was described as constant with progressively increasing intensity. At 9:30 a.m. he was seized by acute severe pain in the left groin that radiated toward the foot. The foot became pale, and shortly thereafter the leg began to tingle, felt numb and heavy, and then became swollen and later turned red and violaceous.

Past history disclosed recurrent pulmonary infection, peptic ulcer, recent weight loss, redness, soreness, and burning of his tongue, and cheliosis of the lips. Besides alcohol, the patient had survived on only eggnog and soups.

Physical examination revealed an emaciated man with slurred, thick speech. The blood pressure was 90/70, the pulse was 140. The tongue was beefy red with infected oral and buccal mucosa and cheliosis. The lungs were normal, a grade-II rough, short apical systolic murmur was present, and the liver edge was enlarged but nontender.

The left lower extremity was diffusely violaceous from the toes to the proximal third of the thigh. The left foot was cooler than the right, and the dorsalis pedis and posterior tibial arteries were not palpable. Femoral pulsation was present. The leg was swollen, tense, and of a woody consistency. There was a suggestion of effusion of the left knee with a ballotable patella.

On admission the hematocrit level was 62 per cent and the white blood count was 28,500 with 83 per cent neutrophils. The bromsulfalein retention was 13.6 per cent in 45 minutes.

The admission electrocardiogram showed sinus tachycardia of 140 beats per minute, right axis deviation with clockwise rotation, and prominent P waves in leads II, III, and aVF, which were consistent with acute cor pulmonale. X-ray of the chest showed cavitory and nodular lesions in the upper lobes suggesting pulmonary tuberculosis.

Bed rest with elevation of the left leg, intra-venous fluids, and large doses of vitamins were prescribed. After an epidural block with 1 per cent procaine through a polyethylene catheter, there was good relief of the pain, some increase
in the temperature of the leg, and loss of most of the cyanosis. The skin still appeared erythematous and had a mottled appearance, with a few petechial hemorrhages. Heparin was given intravenously, 75 mg. every 8 hours.

On the following day the left leg was less erythematous and edematous, without cyanosis or gangrene. There were still no palpable pulses on the left side below the femoral artery. Leg pain continued to be relieved by 20 ml. of 1.5 per cent metyrapone every 4 to 6 hours in the epidural catheter. The patient then developed delirium tremens, for which he was given trifluromazine (Vesprin) and intravenous fluids.

Two days after admission the left posterior tibial artery became palpable, and the leg gradually improved. An acute thrombophlebitis of the lumbar veins also developed and cleared, but deep vein tenderness in the left calf and femoral canal persisted.

Active tuberculosis with positive cultures was found and treated with drugs. On July 25, just before his discharge to a tuberculosis hospital, a venogram showed obstruction in the left external iliac vein and prominent collateral circulation. The patient had received intravenous heparin without untoward effects. There was no hemoptysis, in spite of the active tuberculosis. Edema of the leg and abdominal wall had disappeared and only tenderness of the femoral vein remained.

Case 2

A 30-year-old woman of Greek descent was admitted to The Graduate Hospital on December 5, 1959, complaining of vomiting and weakness of 6 weeks' duration. On physical examination three freely movable tender nodes were found over the left supraclavicular area, the largest of which was 1 by 2 cm. The patient's abdomen was moderately distended, and over 400 ml. of gastric residual were found.

X-rays showed an almost completely obstructing lesion involving the antrum and lower third of the stomach, and two penetrating ulcer craters on the lesser curvature. Intermittent gastric suction was employed with feedings of milk and alkali and administration of phenobarbital and anticholinergic agents.

On the third hospital day the patient developed pitting edema of the left hand, which increased to involve the left cervical, thoracic, and upper abdominal areas, and fever to 101 F. appeared.

X-rays of the chest revealed left pleural effusion with marked tissue swelling of the left axilla and left chest wall. A diffuse erythema of the edematous areas appeared, except for the fingers, which were cold, moist, and moderately cyanotic. The radial pulse was normal. A dull pain was experienced in the left arm and fingers. The apparent diagnosis was venous obstruction of axillary, aygous, and internal mammary veins, with thrombophlebitis and associated arterial spasm, phlegmasia cerulea dolens. A left cervical sympathetic block produced a complete Horner syndrome with definite increase in warmth of the forearm and hand. The deep cyanosis was relieved by the block, as was the pain. Heparin was given intravenously, 75 mg. every 8 hours, the extremity was elevated, and elastic bandages were applied. Gastric obstruction persisted and on December 14 a large, left upper quadrant abdominal mass was felt.

By December 15 the edema had decreased slightly, the temperature had decreased and the cyanosis slowly returned in all fingers of the left hand, as far proximal as the middle phalangeal joint. A tingling sensation in the middle and little fingers, and hypesthesia of the distal phalanges occurred. A partial Horner's sign was still present. The ganglion block was repeated, but the cyanosis of the middle and little fingers remained fixed and did not Blanch with pressure. Vesicular blebs developed on the palmar surfaces of the cyanotic fingers. An infrared photograph showed markedly dilated venous collateral circulation of the left upper extremity and chest wall (fig. 1).

The patient's condition deteriorated rapidly. She still retained only small amounts of oral fluids, and she needed supplementary intravenous infusions daily. She continued to have pain in the left arm, especially the fingers. The distal segments of the middle and the little finger had a dry type of gangrene. The edema of the left arm and chest wall had decreased only slightly and she required narcotics for pain. A biopsy of a left scalene node showed metastatic adenocarcinoma. On December 23 radiation of axillary and supraclavicular areas and the stomach was begun. After 4 weeks she had marked resolution of the edema of the left upper extremity and the chest wall.

Six weeks after admission she was allowed to go home. There was no edema, but dry gangrene of the distal phalanges of the middle and little fingers, left hand, was present. She was still vomiting one to two times daily.

The patient was re-admitted to the hospital 1 week later for a short time because of abdominal pain and vomiting. She was markedly emaciated, but the edema had not returned. The radial pulse was palpable, and the middle and little fingers showed dry gangrene of the distal phalanx. She died at home on February 13, 1960. Autopsy was not performed.

Discussion

The clinical manifestations of phlegmasia

Circulation, Volume XXV, January 1962
cerulea dolens consist of edema, pain, discoloration, vascular impairment, and temperature changes. These manifestations may be sudden or gradual over a 3-day period.

Discoloration is the most characteristic sign and may be present at the onset. It is usually described as a deep violaceous color or cyanosis, and may be mottled or marbled. The discoloration usually begins in the distal portion of the limb and extends proximally. It may be relieved by sympathetic block. The discoloration is probably due to increase in reduced hemoglobin in the blood, which is trapped and cannot leave the extremity.

Many cases of phlegmasia cerulea dolens have been incorrectly diagnosed as arterial emboli because of cyanosis, absent pulses, decreased temperature, hypesthesia, and loss of motor power. The most important differential point is the absence of edema and cyanosis in arterial occlusion.

Of the 73 cases of phlegmasia cerulea dolens reported from 1949 to 1960, 12.5 per cent died during the acute episode or shortly afterwards; 23 per cent developed gangrene that was extensive enough in a few cases to require amputation of the entire extremity. The overall progress was much worse in those cases that developed gangrene. Of the 17 patients with gangrene, six, or 35 per cent, died. This is in contrast to the 5 per cent mortality in those that did not develop gangrene (3 of 56). Death in the patients with phlegmasia cerulea dolens was usually due to circulatory collapse or to an underlying condition, as widespread carcinoma.

Pulmonary embolus developed in 18 per cent (13 of 73) of the reported cases, 30 per cent in the patients who developed gangrene, and 14 per cent among the others. The temperature of the extremity was decreased in 88 per cent (36 of 41 cases) and the arterial

Figure 1
An infrared photograph of case 2 showing the swollen left upper extremity. The collateral venous circulation of the left arm and chest wall is shown.
pulses were palpable in only 39 per cent of the cases (17 of 44).

Gangrene as the result of venous occlusion is usually superficial, and the gangrene of the deeper tissues may be less extensive than is suggested by initial observations. Amputations should be avoided initially, because the gangrene will usually demarcate by itself. Usually there is no need for surgery.

The extremity should be elevated, and the patient given analgesics and sedatives for pain and apprehension and antibiotics for gangrene. Blood, fluids, and vasopressor agents may be given for circulatory collapse if necessary. Sympathetic blocks should be done before anticoagulant therapy is started, because of the danger of bleeding. In several of the reported cases no relief of pain was noted after sympathetic block, and several progressed to gangrene despite sympathetic blocks, as did our second case. We have found continuous caudal block to be most reliable.

For anticoagulation we prefer the use of intravenous heparin. The dose varies according to the size of the patient from 50 to 100 mg. A control coagulation time is taken before therapy is started, and again 24 to 48 hours later just before a dose of heparin is given to determine undue sensitivity. We have found that 75 mg. of aqueous heparin intravenously, every 8 hours, is usually sufficient.

Summary

Two cases of phlegmasia cerulea dolens are presented with acute venous obstruction, arterial spasm, and compromise of circulation to the extremity.

References


Thomas Sydenham
1624-1689

What mattered, considered Sydenham, was that a doctor should come to know diseases, should learn the conditions out of which they arise, should be aware of the means by which they can be cured . . . We have before us a sick and suffering human being. That is our object of study; the symptoms, their changes, the cause of illnesses.

The doctor's business is to assist nature in its struggle, to guide and to intensify the healing power of nature. Now, if the doctor is to intervene to good purpose he must know the particular diseases as well as possible. Particular diseases exist. There are "species morborum," various kinds of illness, just as there are species of animals and plants. Even as the zoologist and the botanist learn how to distinguish animal and vegetable species one from another, so must the physician endeavour to distinguish the various diseases . . . The way to an understanding of particular diseases is their clinical observation from the first symptoms to the disappearance of the last. Sydenham followed up this line with all the zeal of a scientific investigator.—HENRY F. SIGERIST, M.D. The Great Doctors. New York, W. W. Norton & Co., Inc., 1933, p. 179.
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Circulation. 1962;25:39-42
doi: 10.1161/01.CIR.25.1.39

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/25/1/39

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