

Symmetrical peripheral gangrene

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Two cases of symmetrical peripheral gangrene associated with intracardiac lesions are described. One had a secondary deposit from a uterine leiomyosarcoma in the wall of the right ventricle, the other had a free ball thrombus in the right atrium; there were no valvular lesions in either case. The published reports of this type of gangrene indicate that it may result from (1) vasospastic conditions, (2) small vessel obstruction, or (3) conditions producing a very low cardiac output. The two patients reported are unusual examples of the last.

Symmetrical peripheral gangrene is said to be present when the distal parts of two or more extremities undergo ischaemic change without obstruction to the main arteries supplying them (Goodwin and Berne, 1974). The association of this rare condition with an obstructive intracardiac lesion was first described by Von Ziemssen in 1890. This led in subsequent years to case reports of symmetrical peripheral gangrene in which the presence of a left atrial ball thrombus caused by mitral stenosis was suspected clinically and confirmed at necropsy (Bozzolo, 1896; Battistini, 1909; Lutembacher, 1917; Aubertin and Rime, 1926; Covey *et al.*, 1928). In 1931 Schwartz and Biloon described 3 cases of symmetrical peripheral gangrene. Two had left atrial ball thrombi secondary to mitral stenosis and the third had hypertensive heart failure and a left atrial thrombus but without valvular disease.

Symmetrical peripheral gangrene has been described in other conditions in which there is a fall in cardiac output. It has been noted after myocardial infarction (Swan and Henderson, 1951; Cotton and Bedford, 1956; Cohen, 1961), in hypertensive heart failure (Perry and Davie, 1939) and with resistant ventricular tachycardia (Abrahams, 1948).

We present 2 cases of peripheral gangrene associated with intracardiac lesions. In one there was a secondary leiomyosarcoma in the right ventricular wall, and in the other a large ball thrombus of the right atrium. These cases are unusual in that the right side of the heart was affected, and also in that no valvular lesion was found at necropsy.

Case reports

CASE 1

A 53-year-old woman was admitted with cyanosis of her feet and hands. She had had a uterine leiomyosarcoma removed one year previously. On examination she had incipient gangrene of the cheeks, nose, feet, and hands, and demarcation developed as shown (Fig. 1 and 2). She had a sinus tachycardia of 100/min and blood pressure of 135/105 mmHg. The heart was enlarged. Her jugular venous pressure was raised but her peripheral veins were collapsed so that venepuncture was difficult. Both radial pulses were palpable, but no pulses were felt below the femorals. There was a diastolic murmur at the left sternal border and a loud systolic murmur in the pulmonary area. She died one month after admission.

At necropsy a secondary deposit of leiomyosarcoma removed one year previously. On examination she had incipient gangrene of the cheeks, nose, feet, and hands, and demarcation developed as shown (Fig. 1 and 2). She had a sinus tachycardia of 100/min and blood pressure of 135/105 mmHg. The heart was enlarged. Her jugular venous pressure was raised but her peripheral veins were collapsed so that venepuncture was difficult. Both radial pulses were palpable, but no pulses were felt below the femorals. There was a diastolic murmur at the left sternal border and a loud systolic murmur in the pulmonary area. She died one month after admission.

At necropsy a secondary deposit of leiomyo-

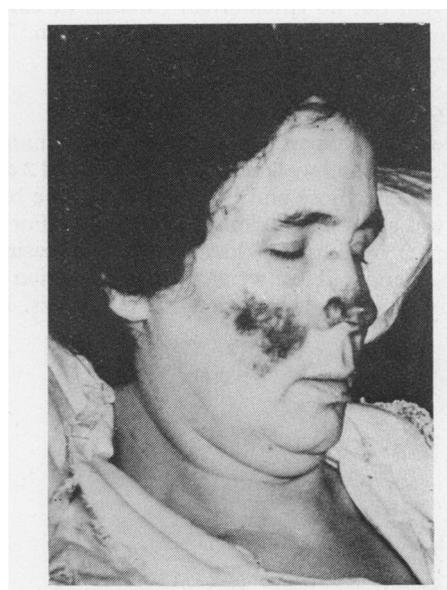


Fig. 1 Ischaemic involvement of nose and cheeks in case 1.

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Fig. 2 *Gangrene of fingers in case 1.*

sarcoma was found in the anterior wall of the right ventricle almost obliterating the cavity. Two secondary deposits were present in the lungs. There was no evidence of local recurrence. No evidence of arterial thrombosis or embolus was found in the femoral, popliteal, dorsalis pedis, or ulnar vessels on macroscopical or histological examination. Recent antemortem thrombus was found in the portal vein but no antemortem thrombi or emboli were found elsewhere in the venous system. The coronary arteries were normal.

CASE 2

A 71-year-old woman was admitted complaining of breathlessness and weakness. She had noticed 2 days before that her nose and cheeks had become blue. There was a past history only of bronchitis. For the previous 2 months she had become increasingly dyspnoeic and had lost weight. On examination she had intense cyanosis of her nose and cheeks. Her



Fig. 3 *Concentric area of ischaemia and ulceration around the perineum in case 2.*

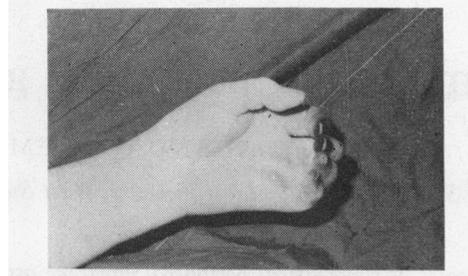


Fig. 4 *Gangrene of fingers in case 2.*

left hand was cold and the nail beds of the second, third, fourth, and fifth fingers were cyanosed. Both feet were cold and slightly cyanosed. There was a concentric area of ischaemia and ulceration around her perineum (Fig. 3). She had a deep venous thrombosis of her right leg. Her blood pressure was 120/80 mmHg. Her pulse was 80 and fibrillating. Her jugular venous pressure was not raised. Both radial pulses were palpable though the left was weaker. No pulses were felt in her legs. On admission haemoglobin 13.6 g/dl, platelets $54 \times 10^9/l$, white blood count $17.2 \times 10^9/l$, 85 per cent polymorphs. Fibrinogen degradation products greater than 80 and less than 160. Treatment was started with intravenous heparin, but she continued to deteriorate and gangrene of her fingers developed (Fig. 4). She died 48 hours after admission.

At necropsy there was moderate atheroma of the aorta. A thin plaque of antemortem thrombus was adherent to the posterior wall of the abdominal aorta and there was a similar thrombus in the left common iliac. There was a deep venous thrombosis of the right femoral vein extending to the right common iliac vein. Both pulmonary arteries were almost obliterated by adherent antemortem thrombus of some days' duration. There was a large free ball thrombus, 43 mm \times 37 mm in the right atrial appendix and numerous small warty thrombi were adherent to the right atrial and ventricular walls (Fig. 5). One similar adherent thrombus was found in the left atrial appendix. There was no valvular disease. Carcinoma was not found.

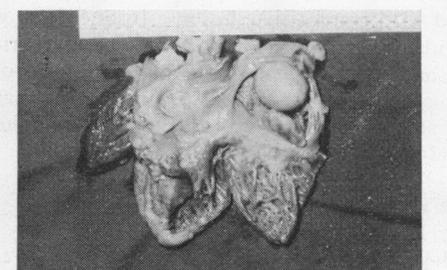


Fig. 5 *Right atrial thrombus and small adherent thrombi in case 2.*

Discussion

In addition to the peripheral limb ischaemia found in symmetrical peripheral gangrene, the nose, cheeks, ear lobes, and occasionally the perineum may also be involved. The causes of this syndrome fall into three main groups: (1) vasospastic conditions such as Raynaud's syndrome (Raynaud, 1888), ergot poisoning (Cranley *et al.*, 1963), and frostbite (Dunning, 1964); (2) conditions in which the small vessels are obstructed, such as disseminated intravascular coagulation (Goodwin and Berne, 1974), the presence of cold agglutinins, or primary thrombocythaemia (Preston *et al.*, 1974) and Lloyd *et al.* (1967) described 4 cases in children in which the presumptive diagnosis was polyarteritis nodosa; it has also been described in children after infectious diseases such as chickenpox (Gyde and Beales, 1970) and measles (Chaudhuri and McKenzie, 1970) where intravascular coagulation was probably responsible; and (3) conditions leading to a fall in cardiac output—our cases belong to this third group.

Peripheral gangrene of the limbs alone is rarely a consequence of extensive venous obstruction caused by thrombosis and occasionally more than one limb may be involved (Ross *et al.*, 1961).

The possibility of diagnosing clinically an occluding atrial thrombus was first suggested by Von Ziemssen in 1890, when he described circumscribed gangrene of the feet with absent or diminished arterial pulsation in the legs in 3 patients found to have large left atrial thrombi and mitral stenosis. The association of symmetrical peripheral gangrene with myocardial infarction, hypertensive heart disease, and arrhythmia has since made it clear that the fall in cardiac output responsible may simply be a result of failure of the heart as a pump, without actual obstruction to flow.

In 1938 Fishberg suggested that the cause of the gangrene was intense peripheral vasoconstriction in an attempt to maintain the blood pressure and circulation to vital organs in the presence of a grossly reduced cardiac output. In his patients he found that, while the neck veins were engorged, the peripheral veins were collapsed and venepuncture was difficult, suggesting poor peripheral flow. He drew attention to the fact that in severe right heart failure the cervical veins were often more engorged than the peripheral veins, presumably a lesser manifestation of this. Fishberg also mentions (1944) that he has seen symmetrical peripheral gangrene in tight mitral stenosis without ball-valve impaction.

Necropsy of the peripheral blood vessels of these patients has consistently failed to reveal thrombosis or emboli, and it has been shown by Roddie and

Shepherd (1957) that the blood flow through human digital arteries falls to zero when the perfusion pressure may still be 36 to 60 mmHg.

Intense cyanosis of the nail beds and cold peripheries is occasionally seen in patients with severe myocardial infarction, but such a dramatic fall in cardiac output is associated with a high mortality, and these patients usually die before frank peripheral gangrene can develop. In our first case the coronary arteries were normal and peripheral vasoconstriction enabled her to maintain an adequate blood flow to her vital organs as the obstruction in her right ventricle increased.

In case 2 the thrombus present in both pulmonary arteries was at least as responsible for the fall in cardiac output as the ball thrombus in the right atrium, and the condition has been described after pulmonary embolus alone (Hejtmancik and Bruce, 1953). There was haematological evidence of intravascular coagulation, and multiple thrombi were found at necropsy, but the distribution of the peripheral ischaemia, particularly of the perineum, suggests that this was the result of vasoconstriction rather than intravascular coagulation. It is in keeping with Fishberg's proposed aetiology that both our patients maintained a reasonable blood pressure while the peripheral gangrene progressed.

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