ACUTE LEFT UPPER LIMB GANGRENE: A CASE REPORT

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ABSTRACT

Background

Acute upper limb gangrene (AULI) is an unusual manifestation of atraumatic acute limb ischemia (ALI), as the latter tends to occur much more commonly in the lower limbs than in the upper extremities. AULI carries a strong risk of limb loss if treatment is delayed or is unsuccessful. Its association with varied co-morbidities further lends to its potential for severe morbidity and mortality. Reported cases worldwide remain few, the dearth of information on the subject being further compounded by its rarity in this environment.

Aim:

This is to report a case of acute left upper limb gangrene.

Conclusion

Early recognition of the features of acute limb ischemia and early prompt medical (anticoagulant therapy) and/or surgical invention yields good long-term outcome, thereby preventing the complications or total loss of limb. A series of successful treatments will demystify the perceived 'spiritual' origin of these conditions.

Key words: Acute upper limb ischemia, acute upper limb gangrene, Limb loss.

Declaration of interest: None.

INTRODUCTION

Acute upper limb ischemia (AULI) is a vascular emergency characterized by sudden occlusion of the arterial supply to the upper limb that threatens the survival of the limb. [1] It is an unusual presentation of limb ischemia, and has an incidence of approximately 3 cases per 100,000 persons per year,[1] one-fifth of the incidence of acute lower limb ischemia. [2, 3] AULI appears to be less common presumably because of the richer collateral supply in the upper limbs than in the lower extremities. [2] Although all age groups can be affected, the elderly are worse hit (mean age of 74 years), [3, 5] The male to female ratio is 2:1. [4] In contrast with our patient, the right upper extremity was found to be more commonly involved for reasons yet unknown. [6] Limb saving treatment should be promptly initiated to re-establish blood flow within 6 hours from onset of symptoms to prevent loss of limb and its accompanying disability. [7]

CASE REPORT

Mrs O.C, a 62 year-old seamstress presented with a 3-day history of moderate dull left hand pain which was noticed on waking up. It radiated to the left shoulder, and was associated with weakness and pale discolouration of the hand which turned dark a day later and progressed upwards to involve the lower forearm. There was no associated fever, malaise, weight loss, nausea or vomiting. She had no prior history of trauma to any of her limbs. She denies ever having used tobacco products. There were no history of her coughing or wheezing, neither was there a history of paroxysmal nocturnal dyspnoea, orthopnoea, intermittent claudication

or limb rest pain. She denied any history of chest pains, headaches, seizures, neck pains or neck stiffness. There was no past history of visual impairment, transient ischemic attacks or strokes. She had a past history of livedo reticularis (a net or lace-like purplish greyish skin macular lesions) on her trunk usually observed when exposed to cold, particularly in early hours of the morning. She denied previous history of any form of haemolysis or anaemia. There were no other known co-morbidities such as Diabetes mellitus, Hypertension or Asthma. Her genotype was HbAA. There was no previous history of similar complaint in her or her family.

On examination she was acutely ill looking but afebrile, with temperature of 36.8^0C. She was observed to be anicteric, not cyanosed, not pale, dehydrated, had no finger clubbing, peripheral lymphadenopathy, pedal oedema, nor stigmata of chronic liver disease. Her waist circumference (WC) was 38 cm and hip circumference (HC) was 43 cm, with a WC: HC of 0.88. There was dark discoloration of her left upper limb extending from the hand to the lower one third of the forearm, which was poikilothermic and wrinkled, with loss of sensation. The brachial, radial and ulnar pulses were absent. The right radial pulse rate was 120/minute, regular, full with no radio-femoral delay. The brachial blood pressure was 130/80mmHg on the right and 0/0mmHg on the left, in sitting position. The jugular venous pressure was not raised and apex beat was in the 5^{th} intercostals place at the mid-clavicular space and no heaves or thrills. Heart sounds S1 and S2 only were heard. There were no palpable aortic abdominal pulsations, organomegaly or ascites. Prompt medical interventions were instituted.

Her left upper limb was warmed with a hot water bottle, and she was placed on oral Nifedipine 20mg and pentoxyifylline 0.5mg, IV benzypenicilllin G 1.2mg and IM tetanus toxoid. There were urgent Vascular Surgical and Orthopaedic reviews. She and her relatives

were counselled on the nature of the illness, and alerted to the high probability of early surgical intervention to rescue the unaffected part of the left upper limb.

Investigations were urgently requested: Doppler USS, FBC + ESR, blood film, clotting profile, renal function and uric acid assays, antibodies screening for antinuclear antigen, double stranded DNA and centomere anti mitochondrial antibody. Despite adequate counselling, she declined further medical treatment, stating a conviction that her ailment was "a Spiritual attack and attack by her enemies". She therefore signed against medical advice, declined any further impute from the medical team, and left the hospital to seek spiritual assistance. Several efforts to trace her for follow-up proved futile.

DISCUSSION

Limb ischemia is considered acute if the vascular occlusion occurs within two weeks of the onset of ischemic features. The site of the vascular occlusion determines the extent of the clinical features, while the progression of the tissue ischaemia depends largely on the functionality of the collateral arterial blood supply and the occurrence or not of continued embolic events. [2] Limb saving treatment should be initiated to re-establish blood flow within 6 hours from onset of symptoms (warm ischemic time). Skeletal muscle tissue, the major component of the extremities is particularly susceptible to ischemia. Adjacent tissues such as the skin and nerves are not spared either. [1, 7] Irreversible muscle cell injury has been shown to begin after 3 hours of ischemia, the damage being almost complete by 6 hours. [7]

AULI may affect both the large and small arteries. The brachial artery has been shown to be the most common site (48.6%) of upper limb vascular occlusion. [4] The significant risk factors include tobacco use, diabetes, hypercholesterolemia, hypertension, and advanced age. [5,8] Others are systemic lupus, rheumatoid arthritis, and other autoimmune disorders. [2] Notable sources of emboli include: atrial fibrillation or other dysrhythmias (80%), recent myocardial

infarct (12%), subclavian artery aneurysm (5%), left atrial myxoma (2%), and occasionally, the superficial palmar arch, atherosclerotic plaque from aortic arch or proximal large vessels and thrombosis of subclavian artery aneurysm. [4,5,9,10] None of these risk factor were observed in our index patient, neither was any prescribed investigation carried out to identify the precise site of her vascular occlusion.

Atherosclerosis and thrombosis of proximal large vessels may also present acutely in a setting of chronic limb ischemia (acute on chronic) as in the index patient. Causes of chronic limb ischemia include Raynaud's disease, Burger's disease, atherosclerosis, thoracic outlet syndrome, and inflammatory diseases (giant cell and Takayasu's arteritis). [4]

The clinical features include an acute onset of pain, pallor, pulselessness, poikilothermia, cyanosis, petechiae, paralysis, tenderness, mottled skin, impaired sensation, muscle weakness and frank gangrene may also be observed. [1, 2] Hairless, dry skin may suggest chronicity. Raynaud's disease is characterized by pallor, cyanosis and post ischemic hyperaemia of digits in sequence (tri-phasic colour change), due to vasospasm following exposure to cold or emotional stress. [4] Additionally, symptoms of connective tissue disorders, such as dry eyes and mouth, swallowing difficulties and arthritis may be elicited. [1, 2] Unilateral symptoms suggest an isolated local aetiology, while bilateral symptoms may imply systemic pathologies such as a connective tissue disorder. [2] Kaar et al in a series of 55 patients with upper limb emboli reported that 27% presented ≥48 hours after the onset of symptoms, while 4 patients presented with frank gangrene. [9]

Segmental blood pressure assessment is useful in large vessel occlusion. A blood pressure difference >10mmHg compared to the contralateral limb could be significant. Finger systolic pressure difference of >15mmHg among digits or an absolute pressure <50mmHg suggests significant pathology. [1, 2] Our index patient had a systolic blood pressure difference of

130mmHg between both upper limbs. CT angiograms clearly outline proximal lesions. Conventional angiography demonstrates the distal vessels of the forearm and hand, and is also used to guide balloon angioplasty and stenting. Doppler ultrasound demonstrates the pattern of blood flow. [2, 4]

Screening for a connective tissue disease may be indicated, a positive serologic test prompting a rheumatology referral. Other investigations such as echocardiography and clotting profile may be employed depending on the suspected background aetiology. [4]

The treatment goal is to re-establish pulsatile blood flow. Acute limb ischemia was largely managed conservatively before the invention of the Fogarty catheter. [2] Turner et al, in a series of 17 patients with AULI, reported that all but one (traumatic vascular injury) were treated conservatively initially, using intravenous heparin(15 patients) and warfarin(1patient who had spontaneous resolution of symptoms) anticoagulation.[2] Conservative management is indicated in selected patient populations who have non-traumatic, non- limb threatening vascular occlusion, or those who are medically unfit for surgery [5] Urokinase or streptokinase are used for acute situations unresponsive to heparinization. [2] Additionally, lifelong warfarin anticoagulation has been proposed for all of these patients. [2]

Raynaud's disease is managed with calcium channel blockers, antiplatelet agents and phosphodiesterase inhibitors. Sympathectomy via video-assisted thoracoscopic surgery is indicated if patient is unresponsive to conservative management. [4]

Certain authors advocated that surgery be considered in all cases, nonetheless, the potential complications in these often poly-morbid patients must be borne in mind.[2] Limb-threatening ischemia requires emergency vascular exploration.[10] The incidence of acute upper limb ischaemia amenable to surgical treatment was 1.3 cases per 100,000/year. [6] Embolectomy, which is now increasingly performed under local anaesthesia, has become the gold standard

in treatment of embolic phenomena, followed by anticoagulation with heparin sulphate, then Coumadin for about 3 months. [2, 3]

Re-embolectomy may be required if there is a recurrence. An amputation should not be considered as treatment failure in patients who present late, as it remains a life-saving procedure. [6] Eyers reported 0–18% amputation rates. [3] Other surgical options include bypass with venous grafts, percutaneous catheter direct thrombolysis,

Pentti et al also reported a post embolectomy mortality rate of 11.3% and death occurred within 30 days from diverse causes including recurrent embolism and myocardial infarction. [6, 9] The mortality was linked to patients' general condition. Good functional outcome was seen in 65-94% of embolectomies throughout a mean follow-up time of 50 weeks. [3, 6]

The possible complications include: re-obstruction, residual arm claudication, wound infection, ischaemic contracture, acute renal failure, and bleeding. [4, 9] Poorer outcome is associated with delayed presentation, symptomatic onset >12hours before treatment, [4] inadequate anticoagulation, and in ischaemia caused by local thrombosis. [6, 9]

Reperfusion of the ischemic limb, though the desired outcome is usually accompanied by undesired local responses such as limb swelling resulting from intracellular and extracellular edema which may lead to compartment syndrome, prolonged ischemia and finally, gangrene; or the systemic response to the release of toxic metabolites and free oxygen radicals which results in membrane damage, leading to multiple organ failure and death.[7] Amputation prevents the flow of these toxins into the general circulation, thereby preventing death. [7]

CONCLUSION

Initial conservative treatment should be considered as the first-line treatment for most patients with AULI especially in this environment where the needed and requested

investigations are often unavailable. Surgery, coupled with anticoagulation remains the gold-standard of treatment. [2, 3]A series of successful treatments will demystify the perceived 'spiritual' origin of these conditions.

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