

Symmetrical Peripheral gangrene in two children at the University Teaching Hospital of Butare (CHUB): a report of two cases

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ABSTRACT

Gangrene of the hands and feet is an uncommon presentation in pediatric patients. There are some known etiologies reported in literature; however, in majority of cases, an etiological factor is not identified. Symmetrical Peripheral Gangrene (SPG) is associated with a high rate of amputation of the limbs in the survivors. No specific treatment has shown to constantly prevent progression of the gangrene.

We report two cases of children with gangrene of the extremities: a fourteen year-old girl with gangrene of all four limbs and a twelve year-old boy with bilateral lower limbs gangrene. Both were acute presentations without any known predisposing factors. For the two cases reported herein, the cause was not clear, although we assume limited diagnostic capabilities may have prevented recognition of more unusual diagnoses.

Keywords: Gangrene - Limbs - Pediatric - Symmetrical - Rwanda

RESUME

La gangrène des mains et des pieds est rare chez les enfants. A présent, quelques étiologies ont été rapportées dans la littérature; toutefois, dans la majorité des cas, un facteur étiologique n'est pas identifié. La gangrène périphérique Symétrique (SPG) est associée à un taux élevé d'amputation des membres chez les survivants. Il n'y a pas de traitement spécifique efficace connu pour empêcher la progression de la gangrène. Nous rapportons deux cas d'enfants avec gangrène des extrémités: une fille de quatorze ans avec gangrène des quatre membres et un garçon de douze ans avec gangrène bilatérale des deux membres inférieurs. Chez tous les deux patients, la présentation était aiguë sans aucun facteur prédisposant connu, la cause n'était pas claire, même si nous supposons que nos capacités limitées de diagnostic pourraient nous avoir empêchées de reconnaître ou de déceler des diagnostics inhabituels.

Mots-clés: Gangrène - Membre - pédiatrique - Symétrique - Rwanda

INTRODUCTION

Peripheral gangrene is an uncommon presentation in pediatric patients. The first SPG case was described by Hutchison in 1891; and it presented with symmetrical peripheral ischemia which lead to a gangrene of two or more sites in the absence of large vessel obstruction or vasculitis. Almost always, a low-flow state is present in association with hypercoagulable vasospastic conditions leading to microcirculatory occlusion [1]. The exact incidence is not known and patients of any age can be affected; however the disease remains rare in children. The causes of SPG can be infectious or non-infectious. The commonest noninfectious etiologies of gangrene of the extremities include trauma, atherosclerosis and diabetes mellitus. The other important noninfectious causes are systemic diseases (systemic lupus erythematosus, progressive systemic sclerosis, Henoch-Schonlein purpura, Takayasu arteritis). The infective causes may be bacterial (Pneumococcus, Staphylococcus aureus, Neisseria meningitides, Streptococcus pyogenes, Klebsiella pneumoniae, E. coli, Salmonella, Pseudomonas), viral (HIV, Rubella virus, and Varicella zoster virus) and parasites (P. falciparum) [1][2][3].

Tropical idiopathic lower limb gangrene (TILG) also known as symmetrical gangrene in the African, Idiopathic gangrene in the African or Idiopathic peripheral gangrene of the tropics is another cause of limb gangrene. TILG

was first reported by M. Gelfand in the Africans in 1947. The features of these first cases were gangrene of unknown etiology, which occurred bilaterally and simultaneously. The first sign was edema of both feet accompanied by pain, and associated with fevers. The onset was sudden and mostly occurred in males who were in the second and forth decades of life. Many cases have been reported in East and Central Africa but recently two variants of the disease have been seen in Nigeria [4]. Rare causes of gangrene include heparin-induced thrombocytopenia, hemolytic uremic syndrome, and traditional oral herbal drugs containing ergot [1][3][5]. Numerous laboratory and radiological investigations are required to find the cause of the gangrene. Nevertheless, in many cases, there is no identifiable etiological factor.

SPG is associated with a high rate of amputation of the limbs in the survivors. No specific treatment has been shown to consistently prevent progression or to reverse the gangrene. [1]

PRESENTATION OF CASES

Patient 1

A 14 year-old girl with no relevant past medical history was admitted from the Surgical Emergency Department at CHUB with gangrene of both four limbs. The patient was healthy with no particular medical concern until five days prior to presentation, when she developed spontaneous right ankle pain while walking. A few hours later the pain was followed by foot and ankle swelling, as well as shivering. On day two, the toes of the right foot started to become blackish, and the left foot was soon similarly

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involved. Two days later, this extremely painful process occurred also in both upper limbs; involving the hands and forearms with concomitant ascending development of blisters in both 4 extremities.

On examination, the patient was healthy, had a low-grade fever, but was hemodynamically normal. She had blisters and darkening of the skin from the feet up to the mid-legs, and from hands up to the mid-forearm on right side extending above the elbow on the left side (Figures 1(a-b)). The right and left dorsalis pedis pulses were absent, though popliteal and femoral pulses were present bilaterally. The right and left radial pulses were present but weak. There were islets of skin necroses on both thighs and buttocks.

Investigations were unremarkable; doppler Ultrasound of both lower and upper limbs showed normal flow pattern, and her HIV status was negative. The patient was started on antibiotics and analgesics; and for one week, was treated empirically with steroids, without improvement.

Three weeks later, the gangrene stopped spreading proximally with clear skin demarcations (Figures 2(a-b)); and the patient had severe pain, barely controlled by usual analgesics. Initially, the parents refused amputation of both 04 limbs, but after clear and full explanation of surgical necessity to amputate the gangrenous parts, they consented for the 04 limbs amputations; which were performed, below the knees for the lower limbs and above the elbows for the upper limbs.

Postoperatively, the patient developed severe malnutrition, and died 3 weeks later of severe sepsis.



Figures 1(a). Clinical appearance on admission
Gangrene of the feet and legs;
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Figures 1(b). Clinical appearance on admission
Gangrene of the upper limbs
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Figures 2. Demarcation of the gangrene 3 weeks later
(a) Demarcation of the gangrene of the lower limbs
(b) Demarcation of the gangrene of the upper limbs
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Patient

A previously healthy 12-year-old boy presented to the Surgical Emergency Department at CHUB with bilateral foot and leg gangrene of 10 days duration. Symptoms started with left foot pain, followed by swelling and fever. The right foot and leg were affected on day two of onset. A topical traditional herbal preparation - which was not specified - was applied to the swollen feet and legs. Two days later, the patient noted progressive darkening of the left toes, with gradual spread to the foot. Blisters developed concomitantly on both lower limbs and were associated with severe pain. In this case, there was also painful buccal mucosal ulceration. In the history, we noted that the patient's brother died 4 years ago with a similar clinical picture at age five.

On clinical examination, the patient had a normal blood pressure, although tachycardic and with moderate grade fever of 38,7oC. The torso was normal, but the lower extremities were dark with blisters and edema of the legs and feet. The left foot was swollen with dry gangrene up to metatarsophalangeal joints (Figures 3(a-b)). The left dorsalis pedis pulse was absent. The right dorsalis pedis pulse was present but weak; and popliteal and femoral pulses were present bilaterally. Investigations were normal; doppler ultrasound of both lower limbs showed normal flow pattern, and his HIV status was negative. The patient was started on broad-spectrum antibiotics and analgesics. The family consented for surgery after multiple attempts for explanation of indication of both lower limbs amputations, and the patient was discharged home 03 weeks after surgery.



Figures 4(a-b). Demarcation of the gangrene 2 weeks later
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Figures 3(a-b). Clinical appearances of the gangrene of feet and legs
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A Written consent formed was obtained from the patients' parents for publication of this case report and accompanying images.

DISCUSSION

SPG in children is rare and the etiology is usually unknown. The exact incidence of SPG is not known. Patients of any age group can be affected. SPG is a well-documented but rare clinical syndrome characterized by symmetrical distal ischemic damage leading to gangrene of two or more sites in the absence of large vessel obstruction or vasculitis [1].

Common etiologies of gangrene of the extremities include trauma, atherosclerosis and diabetes mellitus. However, other important causes of gangrene of the extremities include systemic lupus erythematosus, progressive systemic sclerosis, Henoch-Schonlein purpura, anti-neutrophil cytoplasmic antibody associated vasculitis, Takayasu arteritis, infective endocarditis, and gangrene associated with procoagulant states due to malignancy, anticardiolipin antibody syndrome, and disseminated intravascular coagulation. Rare causes of gangrene include heparin-induced thrombocytopenia, hemolytic uremic syndrome and HIV infection [1, 2, 3].

Noyez et al. described a condition called idiopathic

peripheral gangrene in 12 otherwise healthy Zimbabwean children. A traditional treatment with oral herbs, scarifications or inhalation was considered to be the causative factor of the peripheral gangrene in all of these cases [6]. Similarly, Martins et al. reported four patients with documented *Plasmodium falciparum* infection who developed peripheral gangrene in Maputo, Mozambique [2]; and there has been also another case report of quadruple gangrene of the extremities associated with HIV infection [3].

SPG is associated with a high rate of amputation (auto-amputation or surgical amputation) of the limbs in the survivors. No specific treatment has been shown to consistently prevent progression or to reverse the gangrene [1].

CONCLUSION

Isolated symmetrical peripheral gangrene is a rare condition with multiple possible etiologies. In the two cases we report here, the cause was not clear, although limited diagnostic capabilities may have prevented recognition of more unusual diagnoses.

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