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SYMMETRICAL PERIPHERAL GANGERENE : A CASE REPORT

Surgery

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ABSTRACT

Aims and Objective : "Reporting a case of a 63 year old female presenting with distal gangrene of all four limbs."

Background : Symmetrical peripheral gangrene is a rare clinical syndrome characterized by bilateral distal ischemic damage leading to gangrene of two or more sites in the absence of large vessel obstruction or vasculitis. SPG results in a high mortality rate (upto 40%).About half of the patients who survives requires amputation of the affected limbs.

Methods : A prospective study was done in RIMS, Ranchi, Department of General Surgery in a 63 year old female patient who presented with gangrene of all the four limbs taking into consideration the chief complains , clinical features , lab reports and colour doppler study.

Result : Bilateral lower limb amputation with dysarticulation of the digits of bilateral upper limb was done and post operative phase was uneventful.

Conclusion : Gangrene of all four limbs without the evidence of large vessel obstruction or vasculitis should raise the suspicion of any prevailing cardiac co-morbidity that should prompt early intervention into the cause (like anticoagulation therapy) to prevent the progression of the disease and halt the disease process in the initial stages , thus leading to limb salvaging therapy.

KEYWORDS

Gangrene , Peripheral Gangrene, Amputation, Dysarticulation

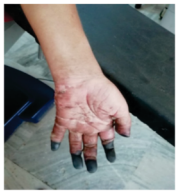
INTRODUCTION :

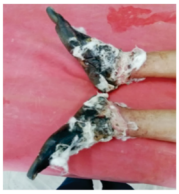
Symmetrical peripheral gangrene (SPG) is a well-documented but rare clinical syndrome characterized by symmetrical distal ischemic damage leading to gangrene of two or more sites in the absence of large vessel obstruction or vasculitis [1],[2],[3] . Symmetrical peripheral gangrene was first described by Hutchison in 1891 in a 37-year-old male who developed gangrene of fingers, toes, and ear lobules after shock. [4]It is a devastating syndrome and mortality is estimated to be up to 40%. More than half of the patients who survive require amputation of the affected limb. [7],[8] Symmetrical peripheral gangrene is a rare syndrome. A recent literature search revealed less than 100 cases. This accounts for the lack of consistent information regarding its incidence, etiology, prevention, and management. [5],[9],[10] The loss of an extremity may manifest as an extensive and evolving threat to the physical, psychological, and social functioning of the patient. [11],[12] The purpose of this report is to present a recent experience with this syndrome, and to discuss potential risk factors and management of SPG. This additional information will help in the prevention of gangrene, reducing the socioeconomic problems arising from amputation.

CASE REPORT :

A 63 year old female presented with blackening of all the four extremities for 1 week . There was a line of demarcation in bilateral lower limbs at the time of presentation. There was no history of trauma, any vascular disease, addiction to tobacco or smoking or use of oral contraceptive pills. Her pre-operative investigation revealed haemoglobin level of 10.7 g/dl, leucocytosis of 21,600/cu mm with platelet count , normal glucose level, normal renal function test, electrolytes within normal range, liver function test revealing a high value of GGT (815 U/L) and raised SGOT (156U/L) and alkaline phosphatase(1111U/L). 2D ECHO was done which revealed global hypokinesia and left ventricular ejection fraction of 25 %.Colour Doppler study suggested normal triphasic flow in the radial and ulnar artey till mid forearm while biphasic pattern distal to it in the right limb while left limb blood flow was normal throughout. Bilateral lower limb blood flow was also normal .

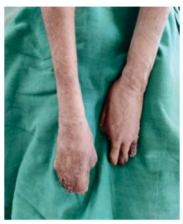
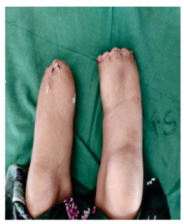
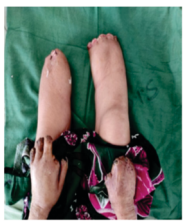
Her Pre Operative Images Are:





Below Knee amputation of bilateral lower limbs and dysarticulation of the digits was performed after taking proper consent.

Her Post Operative Images Are:

DISCUSSION :

The etio-pathogenesis of SPG is not well understood, but the hallmark is the occurrence of microcirculatory failure. [4] It is thus reasonable to speculate that for SPG to become established there must be a low-flow state in the microcirculation of the affected parts, leading to occlusion of these vessels. [5] DIC has been suggested as crucial in the pathogenesis of vascular occlusion. It has been described in several reports as the cutaneous marker of DIC. [4],[6],[8],[13]

Gram-negative septicemia is most commonly associated with clinically overt DIC occurring in 30-50% of patients, and contrary to the common belief, it appears to be equally common in Gram-positive sepsis. [13] Microbial endotoxins (lipopolysaccharide) and exotoxins induce generalized inflammatory response that is characterized by activation of cytokines. The ensuing septicemic shock may lead to occlusion of the reticulo-endothelial system and prevent the rapid clearing of platelet and fibrin microemboli. [14]

Symmetrical peripheral gangrene has also been reported in falciparum malaria. [14] Again, the exact mechanism, which may initiate intravascular coagulation in malaria remains unclear. However, significant parasitemia may activate the complement system, trigger the coagulation pathway, and lead to an alteration in lipid distribution across the parasitized erythrocytes, with an activation of the intrinsic coagulation pathway. The parasitized erythrocytes remain in the microcirculation by a molecular interaction with endothelial receptors, mainly vascular cell adhesion molecule I, intercellular adhesion molecule I, and histidine-rich protein. [7] The sequestration of parasitized erythrocytes due to the decreased deformability of red cells and/or adherence of infected red cells to microvascular endothelium is

thought to initiate microcirculatory occlusion in malaria. [7] Elevated levels of fibrin/fibrinogen degradation products reflecting the ongoing fibrinolysis have been documented, even in uncomplicated falciparum malaria. [14] SPG can also occur as a complication of ergotism, measles, protein C or S or antithrombin III deficiency, and malignancy. It is aggravated by increased sympathetic tone, cold injury to extremities, use of vasopressors, diabetes mellitus and renal failure. [6],[13]

Laboratory markers consistent with the diagnosis of DIC include prolongation of prothrombin time (increased international normalized ratio value), platelet count of <100,000/mm ^{3} , and a low fibrinogen concentration. [15] Tests for fibrin-degradation products or d-dimers may be helpful to differentiate DIC from other conditions that are associated with a low platelet count or prolonged clotting times. [16]

Intense reflex vasoconstriction in the digital vessels of the extremities may cause a nonthrombotic occlusion when the intraluminal hydrostatic pressure falls below the critical closing pressure. [14] The onset is typically insidious and dependent upon blood supply; regions of the body with excellent blood supply are relatively protected, while precarious blood supply served by end arteries increases susceptibility. [6] Frequently affected parts of the body therefore, are the toes, feet, fingers, hands ear lobes and penis. [4],[6] The ischemic changes in the limbs begin distally and may progress proximally to involve the entire extremity. [4] Dry gangrene is commonly a result of arterial occlusion associated with limited putrefaction and absence of invasive bacterial proliferation. [6]

Symmetrical peripheral gangrene can develop at any age and in either sex. [14],[17] A strong index of suspicion by the physician is instrumental in establishing the diagnosis. [2] The first sign of vascular compromise maybe fever followed by marked coldness, pallor, cyanosis, pain and restricted mobility of the extremities. [13] Infrequently, clinical features of the underlying condition may be apparent and help in early recognition of the syndrome. Clinical evidence of DIC such as bleeding from multiple sites, multiple organ failure, and severe acidosis may be present initially or develop subsequently. [14] Cyanosis and hemorrhagic bullae may develop in a symmetrical acral distribution over the fingers and toes. [14] These changes are usually associated in the early stages with intact distal pulses. [5] Symptoms progress and if not reversed, manifests as dry gangrene which becomes apparent within the first 12-24 h after the onset of these changes. [4],[5],[14] This advances proximally with a line of demarcation developing in about 14 days. In severe cases, auto-amputation may occur. [14] Autopsy often reveals thrombi concentrated in the small vessels and not the large vessels. [4],[5],[8],[18]

No treatment is universally satisfactory. Rather, treatment should be guided by the underlying disease and the patient's general condition. [4] The guiding principle is a prompt identification and reduction or elimination of aggravating factors. [4],[13] Accepted first-line measures include, an early and aggressive intervention with appropriate antibiotics for sepsis and shock, and cautious anticoagulation if there is evidence of DIC. [6],[13],[19] Management of falciparum malaria should be started immediately with the suspicion of falciparum malaria as the etiology of SPG. [14] Adjuvant therapy with topical nitroglycerine, intravenous epoprostenol, sympathetic blockade, and hyperbaric oxygen have been reported in the management of established SPG, with dismal outcomes. [4],[8],[13]

The findings of this report suggest similarities with the demographic and clinical pattern described in earlier studies. [4],[6],[8],[13],[14],[17] It is seen in all age groups with no particular predilection for gender, with a devastating clinical outcome of symmetrical multiple limb amputations.

Existing data show that DIC was present in up to 85% of patients with SPG. [20] We considered severe malaria and infection leading to sepsis as the predominant cause of DIC in our patients. DIC is one of the World Health Organization (WHO) criteria for severe malaria, and serve as an independent marker of poor prognosis. [21] The WHO defines complicated malaria as those accompanied with one or more of the following clinical or laboratory findings, that is, an impaired level of consciousness, severe anemia, hypoglycemia, acidosis, hyperlactatemia, hyperparasitemia of more than 5%, and renal impairment. [22] In most of the published cases reporting on SPG, parasitemia is either not mentioned or not exceptionally elevated. Our patient had already received a single loading dose of artemether-lumefantrine before the first determination of parasitemia. SPG appears usually within the first 3 days, even with effective anti-malarial therapy, at which time parasitemia is often very significantly reduced. [15],[23]

Disseminated intravascular coagulation due to bacterial infections is the major cause of SPG. [13],[14],[17] Consistent with diagnosis of sepsis, our patient had an infection with evidence of systemic inflammatory response syndrome. However, the blood and urine cultures were negative. Nevertheless, in the present case and in most of the cases reported in the literature, fever followed by other constitutional symptoms, cyanosis, pallor, and pain in the affected limbs, is strongly suggestive of SPG. [16],[22]

REFERENCES

1. McGouran RC, Emmerson GA. Symmetrical peripheral gangrene. Br Heart J 1977;39:569-72
2. Kalajian AH, Turpen KB, Donovan KO, Malone JC, Callen JP. Phenylephrine-induced microvascular occlusion syndrome in a patient with a heterozygous factor V Leiden mutation. Arch Dermatol 2007;143:1314-7.
3. Davis MD, Dy KM, Nelson S. Presentation and outcome of purpura fulminans associated with peripheral gangrene in 12 patients at Mayo Clinic. J Am Acad Dermatol 2007;57:944-56
4. Jaryal A, Raina S, Thakur S, Sontakke T. Symmetrical peripheral gangrene associated with peripartum cardiomyopathy. Indian Dermatol Online J 2013;4:228-30.
5. Knight TT Jr, Gordon SV, Canady J, Rush DS, Browder W. Symmetrical peripheral gangrene: A new presentation of an old disease. Am Surg 2000;66:196-9.
6. Cartier RA 3 rd , Tchanque-Fossuo C, Asuku ME, Price LA, Milner SM. Symmetrical peripheral gangrene. Eplasty 2012;12:ic10.
7. Gupta A, Dwivedi Y, Saxena AK, Joshi K. Symmetrical peripheral gangrene with Plasmodium falciparum malaria. J Nat Sci Biol Med 2013;4:262-4.
8. Parmar MS. Symmetrical peripheral gangrene: A rare but dreadful complication of sepsis. CMAJ 2002;167:1037-8.
9. Davis MD, Dy KM, Nelson S. Presentation and outcome of purpura fulminans associated with peripheral gangrene in 12 patients at Mayo Clinic. J Am Acad Dermatol 2007;57:944-56.
10. Reid VL, McLay J. Symmetrical peripheral gangrene and disseminated intravascular coagulation associated with pelvi-ureteric junction obstruction. BMJ Case Rep 2013;2013.
11. Norlyk A, Martinsen B, Kjaer-Petersen K. Living with clipped wings-patients' experience of losing a leg. Int J Qual Stud Health Well-being 2013;8:21891.
12. Coffey L, Gallagher P, Desmond D. Goal pursuit and goal adjustment as predictors of disability and quality of life among individuals with a lower limb amputation: A prospective study. Arch Phys Med Rehabil 2014;95:244-52.
13. Gandhi V, Sharma R, Raizada A, Dhawan A. Peripheral symmetrical gangrene treated with sildenafil citrate. J Cutan Aesthet Surg 2012;5:57-8.
14. Sharma BD, Kabra SR, Gupta B. Symmetrical peripheral gangrene. Trop Doct 2004;34:2-4.
15. Liechti ME, Zumsteg V, Hatz CF, Herren T. Plasmodium falciparum cerebral malaria complicated by disseminated intravascular coagulation and symmetrical peripheral gangrene: Case report and review. Eur J Clin Microbiol Infect Dis 2003;22:551-4.
16. Levi M, Ten Cate H. Disseminated intravascular coagulation. N Engl J Med 1999;341:586-92.
17. Kim HM, Roh YT, Park IJ. Symmetrical peripheral gangrene of all fingers and toes: Creating a thumb by on-top plasty in a difficult situation. J Hand Surg Eur Vol 2014;39:897-8.
18. Arora N, Gogna A, Kumar D, Puri K. Peripheral gangrene in Plasmodium vivax malaria: A rare manifestation. Trop Doct 2013;43:23-4.
19. Hotchkiss RS, Levy JH, Levi M. Sepsis-induced disseminated intravascular coagulation, symmetrical peripheral gangrene, and amputations. Crit Care Med 2013;41:e290-1.
20. Ghosh SK, Bandyopadhyay D, Ghosh A. Symmetrical peripheral gangrene: A prospective study of 14 consecutive cases in a tertiary-care hospital in eastern India. J Eur Acad Dermatol Venereol 2010;24:214-8.
21. Ghafoor SZ, MacRae EA, Harding KG, Patel GK. Symmetrical peripheral digital gangrene following severe Plasmodium falciparum malaria-induced disseminated intravascular coagulopathy. Int Wound J 2010;7:418-22.
22. Alkizim FO, Matheka D, Mwanda OW. Malaria complicated by gangrene: A case presentation and review. Pan Afr Med J 2011;10:46.
23. Masse E, Hantson P. Plasmodium falciparum malaria complicated by Symmetrical Peripheral Gangrene, Bowel Ischemia, Repeated Candidemia, and Bacteraemia. Case Rep Med 2014; 2014:696725.