Symmetrical Peripheral Gangrene

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Citation

Abstract
Aim: To present the clinical profile and management of symmetrical peripheral gangrene and review the literature available on the subject.

Methods: Ten patients of Symmetrical peripheral gangrene after initial resuscitation and vital supports were managed either conservatively or amputation was planned depending upon individual case.

Results: Of ten patients, nine survived and one died. Three patients among survivors required amputations. Two patients required delayed skin grafting after wound care, dressing and debridement. Four patients recovered with auto amputation of digits.

Conclusion: Presentation of symmetrical peripheral gangrene is usually with septicemic shock and history of dehydration and gastroenteritis are mostly present. Early aggressive conservative therapy may save life of patient.

INTRODUCTION
Symmetrical peripheral gangrene (SPG) is a rare but devastating syndrome involving distal portions of two or more extremities simultaneously (Figure Legend 1). Disseminated intravascular thrombosis and hemorrhagic infarction of skin with uninvolved proximal arteries are hallmark of this condition. Gangrene of the tip of the nose, ear lobules, lips or genitalia may be accompanied in severe cases. Low flow circulation and septicemia play pivotal role in development of symmetrical peripheral gangrene. Septicaemia is rarely associated with peripheral vascular disease, but a predominant role of infective aetiology is present in symmetrical peripheral gangrene. Common organisms involved are pneumococcus, staphylococcus, and streptococcus but gram negative organisms have also been implicated. SPG can also occur as complication of measles, chickenpox, malignancy, ergotism, protein C or S or antithrombin III deficiency. Aggravating factors are diabetes mellitus, increased sympathetic tone, asplenia, immunosuppression, cold injury to extremities, renal failure and use of vasopressors. Cause of vascular occlusion is not exactly defined, but disseminated intravascular coagulation is present in up to 85% patients with SPG. Low flow state and septicemic conditions are almost invariably present. Fever followed by marked coldness, pallor, cyanosis, pain, restricted mobility of extremity should always raise suspicion of SPG. If aggressive and prompt intervention is delayed, frank gangrene may develop. Ischemia starts and manifests from distal extremity and proximal part are unaffected invariably. Vascular compromise in SPG results in erythematous cold extremities and dusky discolouration of skin. Acral cyanosis and hemorrhagic bulla are followed by development of dry gangrene within 24 hours. Despite therapeutic interventions, different studies report mortality up to 40% and amputation rate of 30 to 50%.

In this article, we present the clinical profile and management of ten cases of symmetrical peripheral gangrene treated at our institute during last 5 years.

PATIENTS AND METHODS
Ten (6 female and 4 male) patients with peripheral symmetrical gangrene presented with evidence of ischemia and gangrene of extremities simultaneously involving either side symmetrically (Figures 1, 2 & 3) between May 2000 and April 2005 at a University Hospital of North India. Empirical antibiotic therapy with adequate intravenous fluid resuscitation was started and if needed, inotropic support was initial plan of management. Enoxaparin at a dose of
1 mgm/kg/day was used to inhibit micro vascular thrombus formation. After 3 days treatment with enoxaparin, sodium warfarin was added until INR 2.0 to 2.5 was reached and then enoxaparin was withdrawn. Antibiotics were either modified or continued according to organism isolated and culture & sensitivity report. Mechanical ventilation and dialysis were supplemented for patients if required. Serial assessment was done regarding improvement and modification of treatment was done accordingly. Patient's profile is shown in Table 1. In all cases, the gangrene was dry with shriveled and necrotic distal extremities. Formation of line of demarcation was evident after 1-4 weeks of the start of the treatment.

**Figure 1**
Figure 1: All limb gangrene

**Figure 2**
Figure 2: Upper Limb Gangrene

**Figure 3**
Figure 3: Lower limb Gangrene

After initial evaluation and adequate resuscitation, following investigations were done included; total & differential leukocyte count, hemoglobin, total protein, platelet count, prothrombin time, serum creatinine, blood urea, serum electrolytes, blood and pus culture & sensitivity.

**RESULTS**
Out of ten patients, only one patient died during treatment. Three patients survived with amputation (One bilateral below knee amputation, one bilateral above knee amputation, one with both hands amputation). In other four cases auto amputation of digits occurred without any other complications. Two patients needed debridement and delayed skin grafting.
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Table 1: Profile of patients with symmetrical peripheral gangrene

<table>
<thead>
<tr>
<th>Parameters</th>
<th>N0. of patients (Percentage)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total patients</td>
<td>10 (100)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>4 (40)</td>
</tr>
<tr>
<td>Female</td>
<td>6 (60)</td>
</tr>
<tr>
<td>Age</td>
<td></td>
</tr>
<tr>
<td>Mean (42.5 years)</td>
<td>-</td>
</tr>
<tr>
<td>Range (28-77 years)</td>
<td>-</td>
</tr>
<tr>
<td>Total leucocyte count</td>
<td></td>
</tr>
<tr>
<td>Leucopenia (&lt;4000/mm³)</td>
<td>2 (20)</td>
</tr>
<tr>
<td>Normal Count (4000-11000/mm³)</td>
<td>6 (60)</td>
</tr>
<tr>
<td>Leukocytosis (&gt;11000/mm³)</td>
<td>2 (20)</td>
</tr>
<tr>
<td>Toxic granules</td>
<td>3 (30)</td>
</tr>
<tr>
<td>Hemodynamic status</td>
<td></td>
</tr>
<tr>
<td>Septicemic shock</td>
<td>7 (70)</td>
</tr>
<tr>
<td>Normotensive</td>
<td>3 (30)</td>
</tr>
<tr>
<td>History of</td>
<td></td>
</tr>
<tr>
<td>Gastroenteritis</td>
<td>6 (60)</td>
</tr>
<tr>
<td>Fever</td>
<td>5 (50)</td>
</tr>
<tr>
<td>Seasonal Variation</td>
<td></td>
</tr>
<tr>
<td>During summer</td>
<td>7 (70)</td>
</tr>
<tr>
<td>During winter</td>
<td>3 (30)</td>
</tr>
</tbody>
</table>

In eight different cases microbes (Pneumococcus, Staphylococcus, Meningococcus, and Pseudomonas) could be isolated (Table 2). Organisms were isolated from pus while blood culture was sterile in majority of cases (80%). Most of the patients (70%) presented during summer. Usually gastroenteritis and fever followed by dehydration and septicaemic shock was present in most cases.

Four patients developed complications during course of treatment needing intensive care unit management. Dialysis support was required in two patients as they developed renal failure. One patient died during treatment due to multiple organ dysfunction syndrome (MODS) while other patient with MODS survived. Three patients developed acute respiratory distress syndrome for which ventilator support was given. Multiple organ dysfunction syndrome developed in two patients for which affected organ dysfunctions monitoring was done and accordingly organ support was given with inotropes, dialysis, ventilation, insulin. Mean hospital stay was 27.4 days with range of 6 to 47 days. Patient developing MODS needed prolonged hospitalization. Results of therapy are in Table 3.
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DISCUSSION

Symmetrical peripheral gangrene is a rare clinical entity presenting with simultaneous involvement of two or more distal extremities. Tip of nose, lip, genitalia or ear lobe may be involved in severe cases. Early diagnosis followed by discontinuation and reduction of vasopressor therapy and proper intervention for sepsis and disseminated intravascular coagulation is needed for successful treatment of peripheral symmetrical gangrene. Previous studies indicate that up to 85% of patients with symmetrical peripheral gangrene have associated disseminated intravascular coagulation. In our study DIC was present in 30% cases. Multiple extremity ischemia should always suggest SPG. Possible etiologic factors e.g. sepsis, DIC, low flow states, vasospasm should be corrected aggressively. Aggravating factors e.g. diabetes mellitus, increased sympathetic tone, corticosteroid therapy, immunosuppression, asplenia, cold injury, renal failure, & use of vasopressors should be identified and managed accordingly.

Treatment plan should be individualized depending on the aggressiveness of disease and type of complication as no treatment is universally effective for SPG. In addition to intravenous fluids with inotropic support, antibiotics, anticoagulants are the main other measures. Several other treatments have been tried like sympathetic blockade either by ganglion block or trimethaphan therapy, intravenous nitroprusside therapy, intravenous infusion of prostaglandin, topical nitroglycerine ointment, and local & intravenous infusion of an alpha blocker (phentolamine, chloropromazine). Use of protein-C and hemodiafiltration have also been tried to correct coagulopathy. Patients of SPG with polychaemia rubra vera may show good response to venesection and chemotherapy with phosphorus or busulfan. Thrombocythaemic patients with SPG respond fairly to the treatment with anti-platelet agents such as aspirin and dipyridamole.

In this study, history of fever and gastroenteritis followed by dehydration was a common factor in most patients. Thus low volume as well as low flow status of body with septicemic shock is usually accompanied with SPG. Disseminated intravascular coagulopathy was present in 30% cases which might be a sequelae of dehydration. We tried to correct septicemic shock, coagulopathy and multiple organ failure followed by decision to proceed for either conservative treatment or amputation. Early intervention reduced mortality to 10% which is less than other studies of 40%. Rate of amputation is 30% which is less than other reports of up to 50%. Auto amputation occurred in 40% patients without any other complications. Debridement and delayed skin grafting was a good option in patients with viable muscle and adequate perfusion. Our two cases recovered after dressing, debridement and delayed skin grafting.

CONCLUSION

In conclusion, peripheral symmetrical gangrene is clinical entity infrequently encountered in surgical practice and requires its separate recognition, as it is usually associated with septicemia, low flow state and coagulopathy while other peripheral vascular occlusive disease causing gangrene are usually not associated with septicemia. Early intervention with antibiotics, intravenous fluids, ionotropes (dopamine, dobutamine, noradrenaline), anticoagulants, and reduction or removal of aggravating factors is essential line of treatment. Depending upon non-viability and line of demarcation of distal parts, amputation is required. As majority of cases have dry gangrene, a well formed line of demarcation is either present or likely to form so amputation should only be done after the line of demarcation has been formed because many cases may have auto amputation.

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