EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 4
Issue: 7
329-331
Year: 2017

SYMMETRICAL PERIPHERAL GANGRENE: A CASE REPORT

[*1]Dr. Dhanwantari Shukla, [2]Dr. Ajay Sharma and [3]Dr. Shambhu Dayal Barua

[1]Surgeon, Sramajibi Hospital, Belur (W.B.).
[2,3]Resident, Department of Surgery, Shyam Shah Medical College, Rewa (M.P.).

***Corresponding Author: Dr. Dhanwantari Shukla**
Surgeon, Sramajibi Hospital, Belur (W.B.).

Article Received on 26/04/2017 **Article Revised on 16/05/2017** **Article Accepted on 06/06/2017**

ABSTRACT
Symmetrical peripheral gangrene (SPG) is a rare clinical syndrome of symmetrical gangrene of two or more sites in absence of large vessel obstruction or vasculitis.[1] Most commonly affected areas are distal extremities, tip of nose, scalp, genitalia and margin of pinna.[2] Exact pathogenesis of the condition is yet elusive, but low-flow circulatory states, vasospastic conditions and hypercoagulable states arising out of sepsis, vasoactive drugs, cardiovascular insufficiency, certain malignancies and connective tissue disorders are associated with this deadly condition.[1] DIC (Disseminated intravascular coagulation) is believed to be the final common pathway of pathogenesis in about 85% of cases.[3] Unless suspected at an incipient stage and a meticulous search to identify and eliminate the offending agent is made, the condition may progress rapidly, leading to loss of limbs and frequently lives. A multidisciplinary team consisiting of intensivists, surgeons and dermatologists is required for comprehensive management of such patients during their acute illness and chronic debilitation.[1] Here, we present a case of an otherwise healthy, 4 year old boy who suffered from symmetrical gangrenous changes in all 4 limbs, following a febrile attack of pneumonia.

KEYWORDS: Symmetrical peripheral gangrene, DIC, pneumonia.

INTRODUCTION
Hutchison, in 1891, first described SPG[4] and since then, the condition has been reported from all parts of the world, mostly in the form of case reports and a few small case series. Often, such patients are immunocompromised in the form of asplenia or chronic renal failure or Diabetes or sepsis[5], but individuals with competent immunity also have been reported to suffer from it. The first signs of pallour, coldness and bluish discoloration in the extremities must incite a high degree of suspicion for an impending SPG and a prompt response from the health care delivery team to find out and treat the etiology, rule out the other possibilities and control the damage already incurred. Surgeon plays a vital role in the team attending to such patients by taking care of the gangrenous organ and the amputation stump, following surgery.

CASE REPORT
A 4 year old boy was presented to the Department of Surgery of a tertiary care teaching Hospital in Central India with the complaints of blackish discoloration of the distal most part of all extremities over past 2 months. This change started following an attack of pneumonia, characterized by fever, malaise, expectorant cough, chest pain and dyspnoea. On presentation, the patient did not have any stigmata of pneumonia. He had symmetrical dry gangrenous changes in both lower limbs those involved the entire feet and lower legs. The left sided gangrene had undergone autoamputation. The distal two-third of the right hand was gangrenous while on the left side, the gangrene was limited to the tips of fingers. The interface between gangrenous and normal parts was well demarcated. Ulnar, radial and popliteal pulses were palpable bilaterally. The vital parameters of the patient were normal and the systemic examination of cardiovascular, respiratory and nervous systems and abdomen did not reveal any mentionable abnormality. The patient was subjected to routine tests available at the Institute viz. Complete hemogram including peripheral smear, Liver Function Test, Renal Function Test, Bleeding time & Clotting time, urine for routine & microscopical examination and Chest X-ray. The test results were essentially normal. To determine the level of vascular occlusion, CT angiography of all 4 limbs was performed which revealed a normal caliber in all large and medium sized vessels with intrinsic occlusion of small sized vessels of all 4 limbs, abruptly from the level of gangrene. Investigations directed towards diagnosing DIC could not be done due to financial constraints. The patient was treated with aseptic dressing of his wounds and amputation of the gangrenous parts was planned. But the guardians of the patient did not consent for the

surgery and got him discharged on request for further opinion.

DISCUSSION

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.[6]

It can affect people of any age group, though the issue of gender predilection is unsettled. Davis et al report male preponderance[7], while Ghosh et al & Tiwary et al report the opposite.[8,9] Often, the condition starts as pallour and and progresses to acrocyanosis and frank dry gangrene, in the background of the underlying associated diseases.[10] Most of the cases occur in winter with exposure to cold acting as a catalyst to the rapid progress of the condition.[5]

A wide array of diseases, characterized by low cardiac output or vasospasm or intravascular coagulation can lead to SPG. The commonest etiological factors can be studied as

1. **Infective causes** which include

Bacterial sepsis due to *Pneumococcus*[11], *Staphylococcus aureus*[7], *Neisseria meningitides.*[7] *Streptococcus pyogenes*[8], *Klebsiella pneumoniae*[12], *E. coli*[8] *Salmonella paratyphi*, *Proteus vulgaris*, *Proteus mirabilis*, *Pusturella multocida*[13], *Pseudomonas*[7], *Enterococcus faecalis*[14], *Capnocytophaga*[7], *Mycobacterium tuberculosis*[15],

Parasitic e.g. *Plasmodium falciparum*[16]

Viral e.g. *Rubeola*, *Measles*, *Varicella zoster*, Viral gastroenteritis[13]

2. **Cardiovascular causes**

Myocardial Infarction[13], Cardiac Failure[13], Hypovolemic shock[2], Hypertension[2], Pulmonary embolism[13].

3. **Malignancies** like- Hodgkin's lymphoma[17], Acute Lymphocytic Leukemia & paraneoplastic syndromes.
4. **Vasopressor drugs** like Adrenaline, NorAdrenaline[18] and Dopamine[19]
5. **Connective tissue disorders** e.g. Systemic Lupus Erythematosus[10], Polymyalgia Rheumatica[17]

The pathogenesis of SPG may involve the Schwartzman reaction, bacterial endotoxin release, and platelet plugging in peripheral arterioles due to vascular collapse and DIC.[7]

The investigations in SPG are directed towards identifying the underlying disease, to confirm DIC with Blood lactate level, Fibrin degradation product, D-dimer assay and the level of vascular compromise by Colour Doppler study, Angiography etc. which characteristically shows sparing of large and medium sized vessels.

Management of SPG focuses on aggressive treatment of DIC[3], management of underlying conditions such as broad-spectrum parenteral antibiotics to treat sepsis and elimination of the agents causing it e.g. vasopressor drugs. The gangrene is dealt by amputation with or without skin grafting, followed by prosthesis where applicable.[5]

Symmetrical peripheral gangrene (SPG) is a rare but devastating complication of septicemia, with a high mortality (up to 40%).[3] About half of the patients who survive require amputation of the affected limb.[17]

CONCLUSION

SPG is an extremely rare sequelae of many extremely common diseases and conditions. Its rarity makes the scientific data available about its pathogenesis and prevention insufficient and inconclusive and once developed, it is a devastating disease as it involves loss of limbs and life. Its association with common diseases and conditions calls for a stronger clinical anticipation and a meticulous and systematic approach towards the problem so that it can be managed optimally.

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