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Calciphylaxis: A case report and literature review

Aishwarya Rajeev[1], Roshni PR[1]

Abstract

Necrotising fasciitis is an extremely life-threatening condition, which mainly develop from the necrosis of the subcutaneous fascia and adjacent tissue. It can be caused recurrently from moderate to severe systemic toxicity and be fatal due to lack of medical or surgical treatment. Any absence of particular clinical features can easily result in the under diagnosis such as cellulites or abscess. The infection can be caused by one or more micro-organisms, both aerobic and anaerobic. Calciphylaxis or calcific uremic arteriolopathy (CUA) is a rare limb threatening condition commonly seen in renal disease patients. It is commonly caused by the accumulation of calcium deposits in the subcutaneous tissue and in the small arteries. Hence, we report a case of 50-year-old female with necrotising fasciitis who was diagnosed with a very rare condition of calciphylaxis.

Key words: Necrotising fasciitis, calciphylaxis, superficial fascia, microangiopathy.

Introduction

Necrotising fasciitis (NF) is an unusual infection of the soft tissues marked by the onset of growing inflammation and necrosis from the fascia including muscles, subcutaneous fat, and necrosis of the skin overlying. (1) It has been reported as a rapidly developing necrotising process followed by severe systemic toxicity by the single or more variety of aerobic and anaerobic organisms. (2) NF is a rare, possibly fatal bacterial infection, which can be developed from appearing like a nonvirulent skin to a necrosis of large area, bullae, and ulceration leads to septic shock. Smoking, diabetes mellitus, intravenous drug use, and peripheral vascular disease are the commonly seen risk factors in adults. Warm skin, erythema, tenderness are the only signs of early NF, whereas severe pain is considered to be the classic initial presentation of NF. (3)

The initiation of the diagnosis of necrotising soft tissue infection is not easily detected. Effortlessly the beginning stages are misinterpreted as cellulitis or abscess. (1) The predisposing factor of NF is found to be diabetes and advanced age. (2) Sepsis or multiple organ system failure with or without respiratory distress syndrome is the indefinite cause of death in NF patients. (4) Calciphylaxis is a life and limb threatening characterised by the calcification and ischaemic cutaneous necrosis. It is usually seen in patients having renal failure or secondary to hyperthyroidism, but also may occur in the absence of above conditions. (5) In 1898 Bryant and White was reported first calciphylaxis and explained about cutaneous gangrene and vascular calcification. Selye and collegues coined the term calciphylaxis in 1962 as a systemic hypersensitivity condition developed in metastatic calcification caused by sensitizing agent. (6)

Here we report a case of NF presented with calci phylaxis.

Case presentation

A 50-year-old female patient who was known as a case of type 2 diabetes mellitus presented with com-

[1] Amrita School of Pharmacy, Amrita Vishwa Vidyapeetham, AIMS Health Sciences Campus, Kochi, India

Address for correspondence:
Ms. Roshni PR
Amrita School of Pharmacy, Amrita Vishwa Vidyapeetham, AIMS Health Sciences Campus, Kochi 682041, India
Tel: 9048540788
Email: roshnipr@aims.amrita.edu

plaints of non healing ulcer over left foot with cellulitis. She had a history of infected non healing ulcer over left heel with spreading infection. The lesion occurred from a nail prick injury over left heel. She was diagnosed with peripheral neuropathy, systemic hypertension, hypothyroidism, chronic kidney disease. Her vital signs were stable, but her laboratory parameters showed some abnormalities (**Table 1**). The patient was empirically started with injection of Magnex Forte 1.5 g iv twice a day, injection of meropenem 500 mg once a day, and injection of metronidazole 500 mg 3 time a day according to endocrine protocol of limb threatening infection. After preliminary investigations, preoperative serology and cardiology fitness were conducted and she underwent left heel grade 2 debridement. Her tissue culture and sensitivity showed the features of Candida non-albicans hence culture specific anti-fungals were initiated. For better wound healing she was given vacuum assisted wound closure therapy. Wounds did not heal and gradually spreading in boundaries with slough (**Figures 1, 2, and 3**). Later, her histopathology revealed suggestion of calciphylaxis. Repeated culture and sensitivity reports showed moderate growth of non fermenter (presumptive Acinetobacter species), Enterococcus species, and Klebsiella pneumonia. Yet her condition remained complicated due to septic shock, septicaemia, NF, end stage renal disease, and diabetic nephropathy, and despite of multiple efforts she expired.

Discussion

NF is described as a serious infection, which mainly affects the soft tissues and those results from the necrosis of subcutaneous tissue, muscle, and fascia. Sepsis and bacteraemia initiates subsequently. Impaired cutaneous wound healing and increased susceptibility to infection can affect the course of soft tissue infection in diabetic patients. The diagnosis from the study of bacteriology in patients with NF is of great value in guiding the empirical antibiotic therapy with specific underlying conditions. (7) But lack of cutaneous findings in the early course of disease and the differentiation from some soft tissue infections, like cellulitis and abscess, makes it more challenging. The survival rates of all critical patients may depend on the early recognition and debridement of all necrotic tissues. The broad spectrum of antibiotics are chosen on the basis of culture reports. (8)

Usually NF can be monomicrobial and polymicrobial. The common monomicrobial agent suggesting from the Taiwan study is Klebsiella pneumonia, whereas frequently occuring polymicrobial species are Escherichia coli, Pseudomonas species, and Proteus mirabilis. The prevalence of type 1 polymicrobial infection was found to be 53.8% according to the reports. (7) Her tissue culture showed the growth of Enterococcus and Klebsiella pneumonia.

This was a report of a 50-year-old female with NF along with non healing ulcer over foot with cellulitis. As her biopsy report revealed calcium deposits, she was diagnosed with calciphylaxis. Calciphylaxis is identified by the intensely painful, ischemic skin lesions marked by the occlusion of micro vessels in the subcutaneous adipose tissue and dermis. Poor life expectancy is generally observed in patients diagnosed with calciphylaxis (<1 year). Calciphylaxis being an underrated disease mostly affects patients suffering from end stage renal diseases, who are known to have a high incidence of extra skeletal calcification. (9)

Our patient had hyponatremia and other electrolytes during the hospital admission, which was critically abnormal than the other studies depicting hypernatremic conditions. Electrolyte imbalance is the widely arising presentation in NF.

Calciphylaxis in some serious condition of inflammations and sclerosis are produced, which causes the accumulation of calcium salts that can promote the resistance of a pathogen in a topical injury. (10)

Conclusion

The mortality rates presented with NF has been always high ranging from 6 to 76% of the population. The continued high mortality in patients with NF is caused by the failure to recognise and diagnose the condition from the early period. The conditions that can make prone to NF include peripheral vascular disease, renal failure, malignancy, diabetes mellitus, odontogenic infection, and alcohol abuse. The people with a history of malnutrition, obesity, arteriosclerosis, and advanced age had reported with higher mortality rates. The extremely toxic conditions can develop in those with anxiety, high fever, leukocytosis, altered mental status, tachycardia, and shock.

Calciphylaxis is an extremely debilitating and a fatal disease if progressive. Superficial fascia is the pathological primary site of NF. The increased growth of bacteria in the superficial fascia causes the release of enzymes and toxins to expand between the fascia. Calciphylaxis is a skin ulceration caused by subcutaneous tissue calcification and in small arteries as a complication of hyperparathyroidism in uremic patients. Painful lesions are appeared in calciphylaxis. The clinical features commonly presented with plaques, nodules, livedo, or purpura, and induration.

Table 1. Laboratory parameters of the patients

Lab parameters	Results
White blood cells	34.84 x 10^9/l
Neutrophils	67.5 x 10^9/l
Haemoglobin	7.7 g/dl
Platelet count	562,000/µl
Sodium	129.5 mEq/l
Potassium	4.0 mmol/l
Urea	73.4 mg/dl
C-reactive protein	148.37 mg/l
Serum creatinine	3.62 mg/dl

Figure 1. Right thigh

Figure 2. Right leg

Figure 3. Left leg and thigh

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