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Case Report

A Calciphylaxis in a post cadaveric kidney transplant man- Case report

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Abstract: We describe a case of calciphylaxis in a 54 years old man known Diabetes Type II, Hypertensive and post cadaveric transplant in the year 2000. Patient became hemodialysis dependent after the diagnosis of calciphylaxis was confirmed. Patient was treated with sodium thiosulfate in addition to intensive wound care. The pathology of the calciphylaxis is poorly understood and an effective treatment is not yet well established. Our observation in this case report that sodium thiosulfate failed to show response probably due to the fact that patient has multifactorial risk factors to associate with calciphylaxis before patient deteriorate and passed away four weeks after the diagnosis of calciphylaxis.
Keywords: calciphylaxis, hemodialysis, sodium thiosulfate

INTRODUCTION

Calciphylaxis is rare; however, the mechanism; the mechanism of Calciphylaxis is poorly understood till date and carries a high mortality rate of up to eighty percent. It requires a high clinical suspicion in order to diagnose at the glance. Bryant and white reported it in association with uremia in the year 1898; however 64 years later Selye demonstrated cutaneous calcification in a nephrectomies rats 1961-1962 [1-3]. Then, a decade and half later Gipstein was able to recognize the clinical important of this calciphylaxis in 11 patients with chronic renal failure syndrome [4]. Since then several case reports have been published in association with uremia and hyperthyroidism or both in combination but not limited to these conditions only [5]. The successful treatment with sodium thiosulfate has been published in case reports and bisphosphonates are a promising treatment for calciphylaxis [5-7].

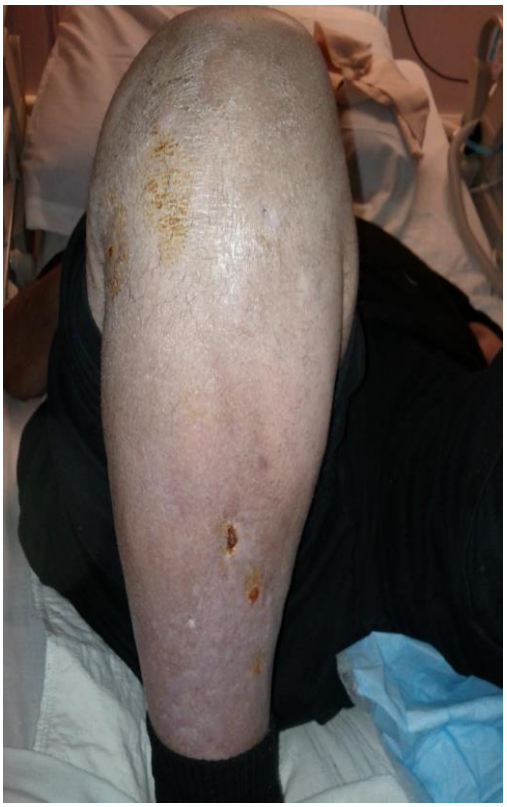
CASE PRESENTATION

54-year-old male known case of Hypertinsion and Diabetes Mellitus Type II on irbesartan and glyburide respectively as he progressed to end stage renal failure on hemodialysis subsequently, he received a cadaveric kidney transplant in the year 2000. Post-transplant patient was on cellcept, cyclosporine, prednisolone and aspirin. In the initial presentation to emergency department he has the right leg redness, swelling and warmness. He was admitted with diagnosis of right leg cellulitis with middle shin ulcer (picture 1). The wound swab revealed pus cells positive, gram staining showed gram-negative rods and culture revealed Serratia marcescens, sensitive to Ceftazidime, Ceftriaxone & Ciprofloxacin. Patient was started empirically on ceftriaxone 2-gram intravenous daily for two weeks and cellulitis was resolved. Further, evaluation of patient in emergency department with right leg ultrasound Doppler showed deep vein thrombosis and was managed with low molecular heparin, however the right shin showed non-healing ulcer which remained after resolution of the cellulitis and deep vein thrombosis. A month later a rapid progression to ESRD required hemodialysis.

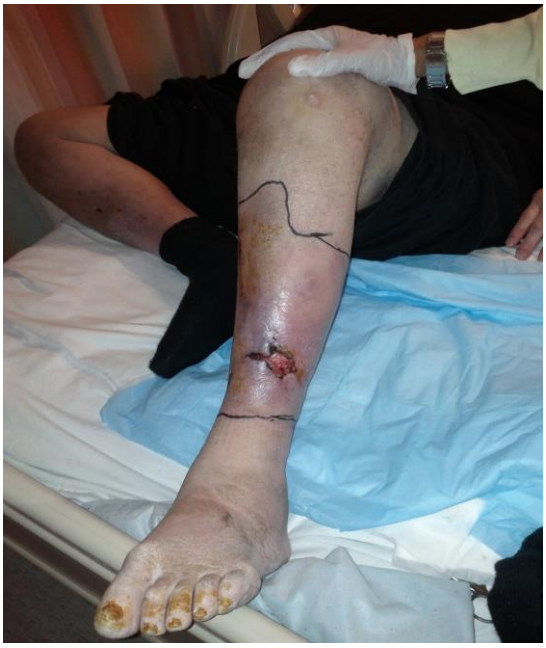
Three months later left shin ulcer with redness, swelling and pain progressing to black necrotic tissue at the edges with alternative spared healthy tissues areas at margins and including the base of the ulcer and similarly, in the medial and anterolateral aspects of left leg where culture and sensitivity showed gram negative rods and culture revealed Serratia marcescens, sensitive to Ceftazidime, Ceftriaxone & Ciprofloxacin as in the (pictures 2,3,4 & 5) and same in the right leg ulcer. He was on ceftriaxone 2 grams intravenously for three weeks and cellulitis resolved. However, patient experienced a progressive severe pain continued in his left leg awakens him from sleep. Necrotizing fasciitis was ruled out. The pain was managed with Tylenol, hydro morphine, fentanyl patch and gabapentin in order to control the pain.

Examination showed an obese male patient in severe pain. Vitals: T 36.5° C, P 73 / minute, 119/96 mmHg , oriented to time place person, jugular venous pressure is 4 centimeter above sternal angle and no hepato-jugular reflex, pale but not jaundiced or

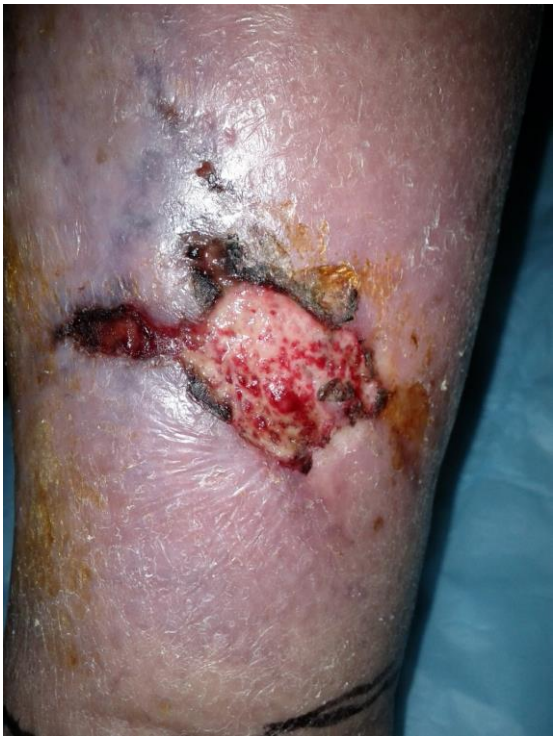
enlarged lymphadenopathies. Chest and CVS, unremarkable, Abdomen examination revealed surgical scars due to kidney transplant in the right iliac fossa. Neurology examination revealed hyperalgesia and pain out of proportion to superficial touch over area of erythema surrounding medial aspect of the right leg lesion, and on medial and anterolateral the left shin and diminished sensation below knee bilaterally for pain, fine touch, temperature, vibration. The peripheral vascular examination revealed posterior tibial, dorsalis pedis, popliteal and femoral pulses are palpable bilaterally but diminished significantly. Locomotors examination showed bilateral non- healing ulcers one on the medial aspect of the right leg shin and three in the left shin medially and anterolateral aspects with redness, warm and tenderness. Investigation showed wbc 16 with predominant neutrophil and hemoglobin 10 gm/dl and normal platelets with normochromic normocytic picture. Random blood sugar 9 mmol/l (normal 4-6 mmol/l, HbA 1C 7.9% (normal 4-6.5%) serum creatinine: 564 mmol/l (80-110mmol/l). Parathyroid hormone, calcium, phosphate, liver function test, PT, PTT, BT, thyroid hormone, protein S, protein C are normal. X-rays of both lower limbs showed calcification of arteries and multiple biopsies of the margin showed a histopathological compatible with picture of calciphylaxis where biopsies showed intimal fibrosis, hyperplasia and calcification within the media of small and medium arteries in additional to vascular micro-thrombi that were seen.



Picture-1: Right Leg Ulcers & Cellulitis (Oct. 6)

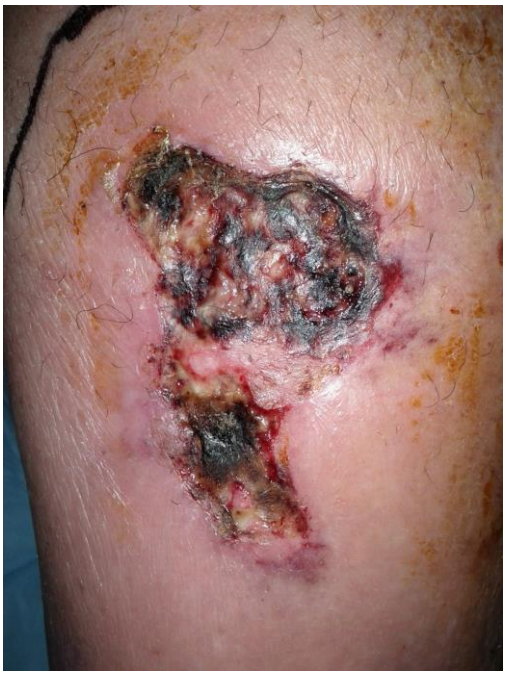


Picture-2: Left Leg Ulcer & Cellulitis - anterolateral aspect (Oct. 6)



Picture-3: Left Leg Ulcer & Cellulitis - anterolateral aspect closer view (Oct. 6)

Picture-4: Left Leg Ulcer & Cellulitis - medial aspect (Oct. 6)



Picture-5: Left Leg Ulcer & Cellulitis - medial aspect closer view (Oct. 6)

DISCUSSION

Calciphylaxis is a rare and serious disorder characterized by systemic medial calcification of the arterioles that leads to ischemia and subcutaneous necrosis. Histologically reveals small vessel mural calcification with or without endovascular fibrosis, extravascular calcification and thrombotic vaso-occlusion leading to ischemic skin necrosis [3, 4].

A little more than half century ago, Hans Selye in the year 1961 identified this finding as calciphylaxis and was able to show that in nephrectomized cases reavealing soft-tissue calcification in rodents and was published in 1962 (1.2).

Calciphylaxis most commonly occurs in patients with end-stage renal disease (ESRD) who are on hemodialysis or who have recently received a renal transplant. It does not exclusively occur in ESRD patients but may be diagnosed in association with primary hyperparathyroidism [3-5].

Treatment successes with sodium sulfate and bisphosphonate have been reported in several case reports. However, it should be continued until the complete resolution of the diseases. Calciphylaxis in a patient with normal renal function have been reported and response to treatment with sodium thiosulfate has been seen. Calciphylaxis due to hyperparathyroidism have been reported and success has been reported post parathyroidectomy [5-7].

CONCLUSION

Calciphylaxis know to be associated with high mortality up to 80 percent of cases. Our patient had multifactorial calciphylaxis associated risk factors that included ESRD, post cadaveric kidney transplant, hemodialysis, DVT and obesity. It is very critical to recognize this diagnosis on initial assessment to confirm the diagnosis and start treatment with sodium sulfate, bisphosphonate or Parathyroidectomy to minimize the high mortality. Unfortunately, our patient passed away four weeks after starting him on sodium sulfate; hence we could not see the outcome in order to confirm the response. However, this case report with visual image may lead to increases awareness of clinicians to appreciate these disfiguring ulcers sooner and use appropriate approach as soon as possible to minimize the high mortality in calciphylaxis associated diseases.

REFERENCES

1. Selye, H., Gabbiani, G., & Strebel, R. (1962). Sensitization to calciphylaxis by endogenous parathyroid hormone. *Endocrinology*, *71*(4), 554-558.
2. Selye, H. (1962). The dermatologic implications of stress and calciphylaxis. *Journal of Investigative Dermatology*, *39*(4), 259-275.
3. Marques, S. A., Kakuda, A. C., Mendaçolli, T. J., Abbade, L. P. F., & Marques, M. E. A. (2013). Calciphylaxis: a rare but potentially fatal event of chronic kidney disease. Case report. *Anais brasileiros de dermatologia*, *88*(6), 44-47.
4. Gipstein, R. M., Coburn, J. W., Adams, D. A., Lee, D. B., Parsa, K. P., Sellers, A., ... & Massry, S. G. (1976). Calciphylaxis in man: a syndrome of tissue necrosis and vascular calcification in 11 patients with chronic renal failure. *Archives of internal medicine*, *136*(11), 1273-1280.

5. Nigwekar, S. U., Kroshinsky, D., Nazarian, R. M., Goverman, J., Malhotra, R., Jackson, V. A., ... & Thadhani, R. I. (2015). Calciphylaxis: risk factors, diagnosis, and treatment. *American Journal of Kidney Diseases*, *66*(1), 133-146.
6. Torregrosa, J. V., Durán, C. E., Barros, X., Blasco, M., Arias, M., Cases, A., & Campistol, J. M. (2012). Successful treatment of calcific uraemic arteriolopathy with bisphosphonates. *Nefrologia*, *32*(3), 329-34.
7. Ross, E. A. (2011). Evolution of treatment strategies for calciphylaxis. *American journal of nephrology*, *34*(5), 460-467.