Calciphylaxis: Challenges in the diagnosis and management

To the Editor,

We read with interest the recent article by Baby *et al.* published in your esteemed journal.[1] With our recent experience, we would like to mention few additional comments which we believe would add to the fund of knowledge of JFMPC readers.[2,3]

Apart from Coumadin-induced skin necrosis, there are a few other close differentials for CUA such as pyoderma gangrenosum, purpura fulminans, antiphospholipid antibody syndrome and endarteritis obliterans. We recently reported a patient with chronic nonhealing penile ulcer who was found to have a penile CUA.[3] Also, Jain *et al.* in their recent report presented an interesting patient who was found to have a synchronous diagnosis of vasculitis and CUA at the same time.[4] Hence, it is very important for the readers to understand the importance of ruling out the other possible differentials in cases with confirmed CUA as more than one disease can co-exist together. Also, to note that there are many other non-ESRD causes of CUA, which we should be aware of like primary hyperparathyroidism, malignancy, Crohn's disease and so on.

Baby *et al.* mentioned the importance of excisional or punch biopsy for adequate biopsy sample to make the diagnosis of CUA. However, skin biopsy is not risk free and there are always chances of nonhealing ulceration, propagation to new lesions, bleeding and necrosis. Hence, we support the practice of avoiding skin biopsy as much as possible especially in cases of ESRD who present with classical dermatological findings of painful ulcerations covered by a black eschar. An expert opinion from dermatologist might be beneficial in cases with complex skin lesions when diagnosis is in question.[5] Deposition of calcium can compromise the organ function slowly and impair the architecture permanently[6].

As mentioned by Baby *et al.*, sodium thiosulfate (STS) stands as a backbone of CUA treatment. Readers must be aware about the commonly reported side effects of STS: High anion gap metabolic acidosis, hypocalcemia, QT interval prolongation and risk of skeletal fractures.

With regard to the recent advancement, a number of newer and experimental therapies have been evaluated in CUA: low-dose infusion of tissue plasminogen activator, hyperbaric oxygen therapy, and low-density lipoprotein apheresis. In addition to coumadin, iron has also been reported as a risk factor for CUA. Hence, if possible, all iron products should be stopped once the diagnosis of CUA is entertained. In ESRD patients on PD, bicarbonate buffered PD fluid is preferable over lactate-buffered fluid, as later has been shown to accentuate vascular calcification.

In conclusion, CUA is a rare calcific disorder and there are significant gaps in understanding the epidemiology, pathophysiology, risk factors and available treatment options. Thereby every attempt should be made to educate the internists, primary care providers, nephrologists and dermatologists to understand CUA, its risk factors, approach to diagnosis and treatment based on the most updated and scientifically proven literature. *European Calciphylaxis Network* (EuCalNet) and *Partners Calciphylaxis Biobank* (PCB) are two international bodies currently working actively to establish an efficient networking system for scientific exchange and collaboration in the area of CUA.

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Conflicts of interest

There are no conflicts of interest.

Kamal Kant Sahu[1], Amos Lal[2], Ajay Kumar Mishra[1], Iryna Chastain[1]

[1]Department of Internal Medicine, Saint Vincent Hospital, Worcester, Massachusetts, [2]Division of Pulmonary and Critical Care Medicine, Mayo Clinic, Rochester, USA

Address for correspondence: Dr. Kamal Kant Sahu, Department of Internal Medicine, Saint Vincent Hospital, 123 Summer Street, Worcester 01608, Massachusetts, United States.
E-mail: drkksahu85@gmail.com

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