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Case Report

Calciphylaxis: The potential diagnostic role of radiologists ☆

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ABSTRACT

Calciphylaxis is a deadly disease that is currently diagnosed through a skin biopsy of resultant necrotic lesions despite many cases lacking this characteristic finding. Recently, research has demonstrated the ability of various radiologic techniques to detect calciphylaxis and have promoted their capabilities in earlier diagnosis without tissue invasion. In this case, an obese 55-year-old female with end stage renal disease, and a long history of dialysis, complained of weeks of persistent abdominal pain that was accompanied by a mottled, lacey, net-like rash that resembled livedo reticularis. Computed tomography of the abdomen revealed extensive arterial calcification and subcutaneous calcium deposition. These radiologic findings, coupled to a high clinical suspicion, prompted treatment for suspected calciphylaxis. Remarkably, after 1 week the patient reported substantial improvement. Hopefully, this publication in conjunction with previous and future research will raise awareness on the role Radiologists can play in expediting the diagnostic process for a lethal disease, especially when a tissue biopsy is not a feasible option.

Introduction

Calciphylaxis, also known as calcific uremic arteriolopathy, is a poorly understood condition characterized by arteriolar calcium deposition with resultant ischemia and possible necrosis of the skin and subcutaneous tissues [1]. The condition is primarily seen in end-stage renal disease patients on hemodialysis and has a reported annual incidence of 0.35% in this population [2]. Other factors shown to have an association include a longer history of dialysis, elevated parathyroid hormone, use of calcium containing binders, diabetes, obesity, and female sex [3]. Although rare, the disease carries a high mortality with an estimated 6-month survival rate of 50% [4]. Currently, a tissue biopsy is recommended to make the diagnosis. However, recent publications [5–7] have shown the ability of computed tomography (CT), X-ray and ultrasound to detect subcutaneous vascular calcifications in order to make the diagnosis of calciphylaxis. Another study found the diagnosis could be made with skeletal scintigraphy through technetium-99m methyl diphosphonate radiotracer uptake in subcutaneous tissues [8]. This case will support previous research and suggest radiologic findings, coupled to a high clinical suspicion, can play a vital role in the prompt recognition

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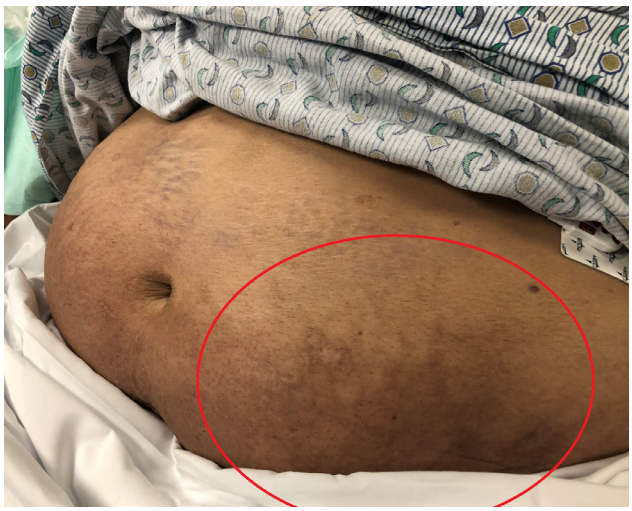

Fig. 1 – Red circle showing a mottled, lacey, net-like rash resembling livedo reticularis in the region of pain.

and diagnosis of calciphylaxis, especially when a characteristic lesion is unavailable for biopsy.

Case report

A 55-year-old obese female with a past medical history of end-stage renal disease requiring hemodialysis via right upper extremity arteriovenous fistula over the past 9 years, secondary hyperparathyroidism, insulin dependent type 2 diabetes mellitus, New York Heart Association Stage 4 diastolic heart failure, atrial fibrillation, and peripheral artery disease presented to the emergency department over concerns of weakness and fatigue after missing over 1 week of hemodialysis sessions

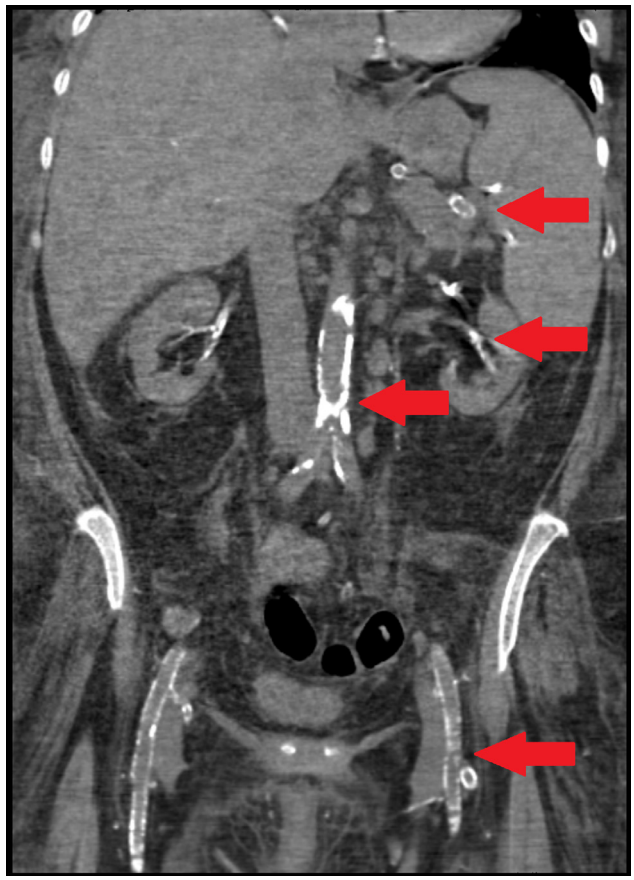

Fig. 2 – CT abdomen coronal view. Superior to inferior the red arrows demonstrate calcification of: splenic artery, bilateral renal arteries, abdominal aorta and branches of the iliac artery.

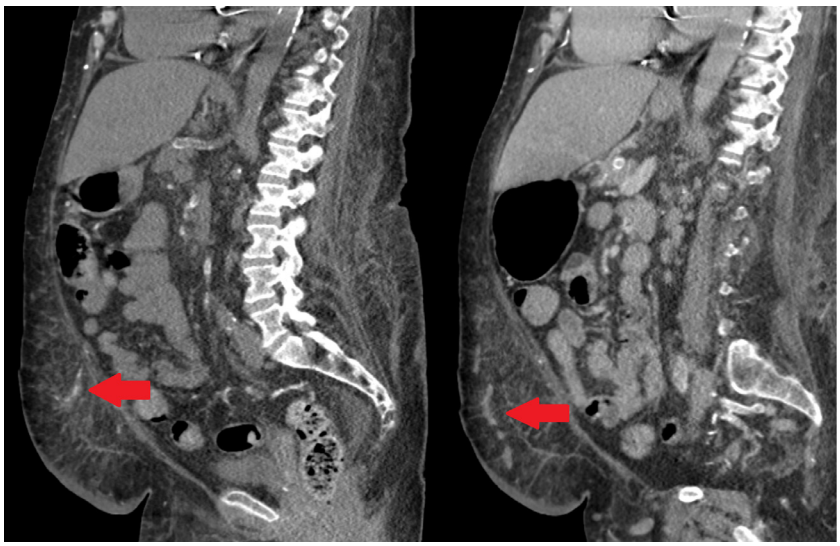

Fig. 3 – CT abdomen showing two sagittal views. Red arrow on the left highlighting subcutaneous calcifications and the right arrow showing subcutaneous soft tissue densities.

that followed a Monday, Wednesday, Friday regimen. On presentation her weight was 246 lbs (BMI 45 kg/m^2) despite her baseline averaging around ˜220 lbs (BMI ˜40 kg/m^2). On examination, her abdomen was distended, and she had pitting edema extending to her thighs bilaterally. Electrocardiogram showed she was in atrial fibrillation with rapid ventricular response. A basic metabolic panel revealed hyperkalemia, so she was given emergent hemodialysis. Soon she developed hypoxemia and severe hypotension with systolic readings as low as 70 mm Hg. She was placed on BiPAP and transferred to the medical intensive care unit where pressor support with Norepinephrine was started. Although she remained afebrile, complete blood count revealed neutrophil predominant leukocytosis and blood cultures grew *Acinetobacter baumannii* of unknown origin. After 1 week, with antibiotics, the patient's infection had resolved and she was weaned off intravenous pressor support. She was transferred to the floors where her fluid status and labile blood pressures were further managed and monitored. While on the floors, the patient's primary complaint was persistent 9-10/10 left lower quadrant abdominal pain. Examination of the area of pain revealed a mottled, lacey, net-like rash that resembled livedo reticularis as shown in Fig. 1. The pain was investigated via CT abdomen that reported anasarca and extensive arterial calcifications (Fig. 2). The imaging also showed lower abdominal subcutaneous soft tissue densities and calcification as shown in Fig. 3. For pain management, the patient was given Oxycodone that would bring her pain down to a 6/10. A short course of steroids was also tried but provided no benefit. Nine days after being transferred to the floors, the patient again developed hypotension in the setting of suspected gastrointestinal bleed supported by hemoptysis, tarry stools, and a positive fecal occult blood test. A CT abdomen with contrast revealed splenic infarction but no signs of active bleeding (Fig. 4). The patient received a couple units of packed red blood cells and fresh frozen plasma. Fortunately, her hemoglobin and hematocrit stabilized, and no further investigative studies were undertaken. Despite persistent abdominal pain, with her fluid status and weight back to baseline, she was discharged to a long-term acute care facility. At the facility, her dialysis sessions were doubled taking place every day apart from Sunday. Despite these efforts, she again began retaining fluid and was re-admitted 6 days after being discharged for heart failure exacerbation. Physical exam showed the rash from previous admission (Fig. 1) and she still complained of 9-10/10 abdominal pain. This pain and rash coupled to the previous radiologic findings (Fig. 2) prompted the decision to withhold her phosphate binder (calcium acetate) and incorporate Cinacalcet to her medicinal regimen for suspected calciphylaxis. After 1 week, the patient reported resolution of her abdominal pain, at which time she was discharged back to long-term acute care.

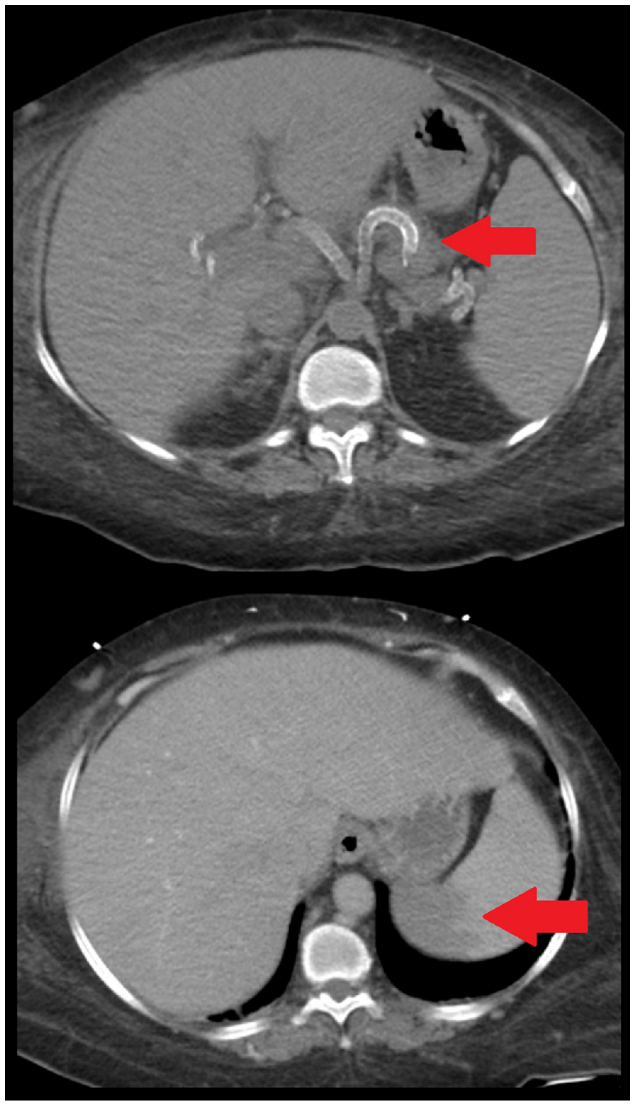

Fig. 4 – CT abdomen with contrast showing two transverse views. Red arrow superior cut showing calcification of the hepatic and splenic artery. Inferior arrow showing consequent splenic infarction.

Discussion

Previous studies have suggested a deep skin biopsy with subcutaneous adipose tissue is required in order to make the diagnosis of calciphylaxis [9]. More recently, the literature seems to be shifting its frame of thought. Prompt recognition of calciphylaxis is crucial for prognosis. It has been shown that approximately 80% of skin manifestations never form the classic indurated necrotic lesion targeted for biopsy [10]. In addition, obtaining a biopsy of viable tissue may serve as a catalyst for the development of extensive necrosis and its subsequent complications [11]. For these reasons, there is interest in exploring more timely, less invasive diagnostic techniques. In this case, the patient had a textbook presentation that included pain in the region of a mottled, lacey, net-like rash that resembled livedo reticularis, arterial and subcutaneous calcium deposition on radiologic imaging, a long history of dialysis, elevated parathyroid hormone, use of calcium containing binders, diabetes, obesity, and female sex. [3] She had an extensive work up investigating other etiologies without success. Although her presentation preached calciphylaxis, she had to suffer from severe 9-10/10 abdominal pain

for weeks before receiving appropriately targeted therapy due to the lack of a necrotic lesion for biopsy – as research suggests is the case for the majority of calciphylaxis patients [10]. Fortunately, clinical suspicion coupled with radiologic evidence was of enough persuasion to treat for calciphylaxis by holding the patients Calcium Acetate and incorporate Cinacalcet to her medicinal regimen. Remarkably, the patient reported improvement in her symptoms just over 1 week later. Hopefully, this publication, in conjunction with previous [5–8] and future research, will raise awareness on the role Radiologists can play in expediting the diagnostic process for a lethal disease, especially when a tissue biopsy is not a feasible option.

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