Calciphylaxis in patient with peritoneal dialysis: A case report

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Calciphylaxis in patient with peritoneal dialysis: A case report

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Abstract: Calciphylaxis is a rare, life-threatening syndrome of vascular calcification, and the pathogenesis of the disease is not well demonstrated. Here, we presented a 33-year-old Chinese male, treated with continuous ambulatory peritoneal dialysis (CAPD) for 5 years with recent onset of calciphylaxis was described. The initial laboratorial workup showed severely elevated intact parathyroid, procalcitonin and serum phosphorus. The main signs were massive pumps in his shoulder and sternum, and necrotic ulcer covered with a black eschar on the right middle finger. The main incentive of calciphylaxis might be the misuse of vitamin-D receptor analogs.

Subjects: Diabetes; Metabolic Disorders; Obesity; General Medicine; Immunology; Nephrology; Dialysis

Keywords: Calciphylaxis; cinacalcet; parathyroidectomy; patient education

1. Introduction
Calciphylaxis is a rare, life-threatening syndrome of vascular calcification characterized by occlusion of microvessels in the subcutaneous adipose tissue and dermis that results in intensely painful, ischemic skin lesions (Nigwekar, Thadhani, Brandenburg, & Ingelfinger, 2018; Nigwekar et al., 2016). Calciphylaxis can be classified into uremic or nonuremic, according to the involvement of end-stage renal disease (ESRD). However, calciphylaxis is most commonly seen in ESRD or hyperparathyroidism, and 70–80% of uremic calciphylaxis with a central distribution (Nigwekar et al., 2018; Weenig, Sewell, Davis, McCarthy, & Pittelkow, 2007). The peripheral lesion tends to...
have a lower body mass index, are more likely to be male and have a lower mortality (Nigwekar et al., 2016; Zitt et al., 2013).

Here, we present a clinical case of uremic calciphylaxis, presented with persistent pain of right fingers for 1 month and exacerbated for 7 days.

2. Case
A 33-year-old Han Nationality, Chinese male, who had been treating with continuous ambulatory peritoneal dialysis (CAPD) for 5 years for ESRD of unknown etiology, complained with a massive lump in his right shoulder for 3 months, darkening and intense pain of right fingers in for 1 months was admitted in department of nephrology. In the recent 1 year, he had not followed in the nephrologists’ outpatient, with intact parathyroid hormone (iPTH) as high as 1700 pg/ml and insufficient peritoneal dialysis. He had lost 10 kg during the 3 months, with the body mass index (BMI) of 18.4 kg/m².

On admission, his blood pressure was 137/78 mmHg, heart rate was 84 beats per minute, respiratory rate was 19 breaths per minutes and body temperature was 36.4°C. The main positive signs were the following. (1). Palpable painless massive nodes on his right shoulder joint and right para-sternum, about 20*15 cm² and 10*10 cm², respectively, and the margin was unclear. (2). Right fingers' tips presented with severe rest pain, and necrotic ulcer partially covered with a black eschar on the right middle finger, without exudation (Figure 1(a,b)). (3). The skin temperature of his limbs was lower than normal with wake pulsation of both dorsalis pedis arteries.

Laboratory results showed a normal hematocrit, platelets, and white blood count. His hemoglobin was 94 g/L (9.4 g/dL), and blood chemistry was in line with ESRD. The corrected plasma calcium was 2.54 mmol/L (10.1 mg/dL), phosphate 2.9 mmol/L (8.9 mg/dL), alkaline phosphatase 548.5 U/L, bone alkaline phosphatase 136.96 μg/L (reference range 11.4–24.6 μg/L), albumin 27.7 g/L (2.77 g/dL), iPTH 3308 pg/mL (350.8 pmol/L), C-reactive protein 137.9 mg/L, procalcitonin

Figure 1. (a, b) Violaceous patch with surrounding retiform purpura, necrotic ulcer; (c,d) H&E stained slides (x40); (e,f) calcification of small vessels and calcific thrombi; (g) necrosis of epidermis.
(PCT) 9.87 ng/ml. Both antineutrophil cytoplasmic antibodies, antinuclear antibodies and complements were negative.

The ultrasound revealed enlargement of the parathyroid, with 1.6*0.7 cm and 1.6*0.9 cm. The Chest computed tomography (CT) revealed diffuse bilateral calcification patches in his shoulder joint and sternum (Figure 2(a,b)). Coronary computed tomography angiogram (CTA) revealed total coronary calcification score was 367.59, with left anterior descending artery scored 362.59, and right coronary artery 4.97.

The patient was subsequently diagnosed with uremic calciphylaxis and received PD+HD (peritoneal dialysis six times per week, except for the hemodialysis day and hemodialysis once a week), intravenous sodium thiosulfate (STS) at a dose of 6 g per day. Meanwhile, prescribed with Cinacalcet 25 mg per day for 14 days (adjusted to 50 mg per day, due to uncontrolled iPTH), and Sevelamer 800 mg three times a day. After 2 months of the prescription, the pumps disappeared and calcification patches were significantly ameliorated (Figure 2(c,d)). The patient felt no pain of his fingers after 7 days.

Once the infection of his digits was controlled, we conducted amputation surgery of his distal part of right middle finger under Multiple Discipline Therapy (MDT). The stitches were removed at day 15 post-surgery (Figure 3). The pathologic diagnosis of the amputated finger confirmed the diagnosis of calciphylaxis (Figure 1(c–g)).

After 55 days of treatment with Cinacalcet and STS, he still presented with severely elevated iPTH, disorder of calcium and phosphorus (Figure 4). The 99m Tc-MIBI scintigraphy revealed parathyroid with 1.1*0.9 cm, 0.8*0.7 cm and 0.4*0.5 cm. Due to the failed control of severe hyperparathyroidism, under the guidance of 2017 Kidney Disease: Improving Global Outcomes (KDIGO) guideline on Chronic kidney disease–mineral and bone disorder (CKD-MBD) (Supplements KI, 2017), the patient underwent parathyroidectomy under MDT, then discontinued Cinacalcet and Sevelamer.

Figure 2. (a) (1st day) calcification patches of right shoulder, 88 mm*148 mm; (b) (1st day) calcification patches of on the right side of sternum (4th thoracic vertebra level); (c,d) (55th day) no calcification patch.
3. Discussion

For most of the cases, uremic calciphylaxis happens in the elder and female with moderate progression, and manifested with central lesion (Brandenburg et al., 2017; McCarthy et al., 2016). In this case, the patient was younger, with a rapid progression, who had several other risk factors for calciphylaxis, including ESRD, hyperphosphatemia, hyperparathyroidism, elevated alkaline phosphatase, hypoalbuminemia, rapid weight loss and medication (VDRAs), etc. But there were several outstanding risk factors for the development of calciphylaxis in the case. First, the patient did not follow-up well, which contributed to the extremely elevated iPTH; Second,
peritoneal equilibration test (PET) indicated insufficient peritoneal dialysis, and the patient refused to undergo hemodialysis alone. The insufficient dialysis deteriorated CKD-MBD. What is more, VDRAs intake under the condition of severe hyperparathyroidism and hyperphosphatemia could be the ignition of calciphylaxis.

The diagnosis was based on his clinical manifestation, and further confirmed by biopsy of the distal part of right middle finger. There were some intriguing conditions that happened during the treatment of the patient. First, after 7 days prescription of hemodialysis-combined peritoneal dialysis, Cinacalcet, Sevelamer and STS, the patient felt no pain in his fingers and the pumps in his shoulder and chest disappeared; Second, the PCT test sustained a high level, and the white blood cell count was under reference range. We treated with antibiotics and the PCT still did not change much; Third, the healing time of wounds of amputation and parathyroidectomy was about 14 days, which mean surgery wound was recoverable without the influence of calciphylaxis.

The treatment of calciphylaxis is comprehensive including wound management, elimination of underlying risk factors and pharmacotherapy. It is important that treatment schema should be in a multidisciplinary manner (Celik et al., 2016; Nigwekar et al., 2018). Under the MDT, we dealt with the wound, and enhanced the management of calcium-phosphorus-iPTH and infection. The combination of MDT and low dose STS was effective to alleviate the ectopic calcification and relief the pain. Here, cinacalcet was effective to shrink the proliferated parathyroid, but it failed to control the level of iPTH. Combined PD and HD has gained popularity in some Asian countries, especially in Japan, where about 20% of all patient received the module (Maruyama & Yokoyama, 2016; Nakai et al., 2014). Compared with PD alone, PD+HD could improve inadequate dialysis and fluid overload, and has the potential effects on improving quality of life and correction of peritoneal deterioration (Maruyama & Yokoyama, 2016). Because the patient refused to treatment with hemodialysis alone, we chose PD+HD for him.

In summary, we presented a case of young male PD patient with ulcerative calciphylaxis. The failure of CKD-MBD management and misuse of VDRAs might be responsible for the development of calciphylaxis. The management of calciphylaxis is MDT and early intervention, which could reverse the disease progression; meanwhile, personalized therapy should be applied. Combination of PD and HD might be a choice for PD patient who reluctant to hemodialysis alone.

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