

Calciophylaxis - A CASE STUDY

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Introduction:

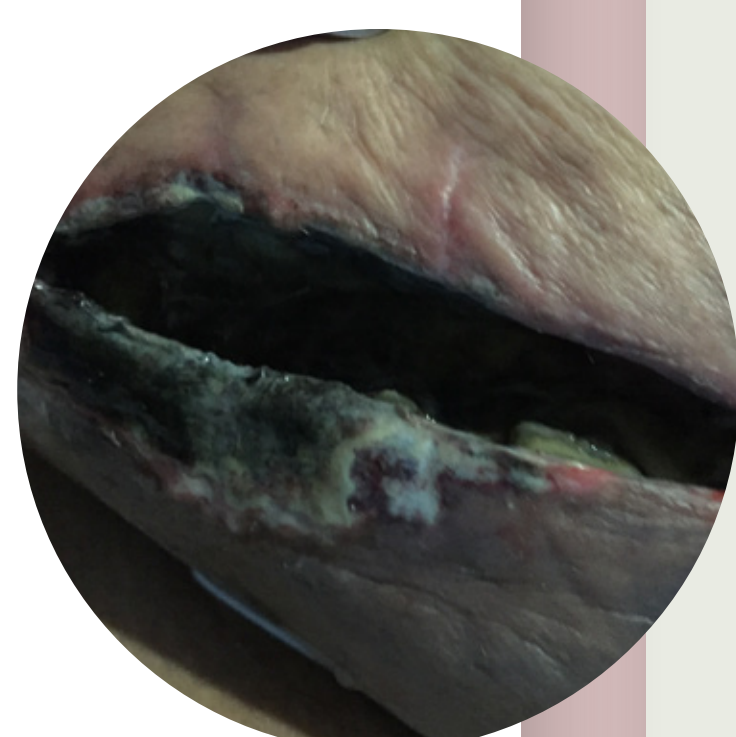
- A rare, life-threatening syndrome of vascular calcification causing occlusion of microvessels in subcutaneous adipose tissue and dermis
- Intensely painful, ischaemic ulcerations and often tactile hyperesthesia
- Rapidly accelerates to starlike, malodorous ulcers with black eschars
- Quality of life affected by ongoing pain, anorexia, insomnia, depression.
- Wound sepsis is most common cause of death
- Survival generally < 1year.
- Ethnicity does *not pre-dispose* to Calciophylaxis
- Predisposition for abdominal/ thighs fat pads - adipose tissue and dermis

Patient Profile

- 47 year old, ATSIC, female
- Morbidly obese - BMI 50.9
- Type 2 DM x 20 years – R insulin
- TIA/stroke – 16 years ago
- Secondary hyperparathyroidism
- ESRD – haemodialysis 5 years
- **Non-compliance of medication and dialysis**
- HTN, dislipidemia, gout
- Grade 1 uterine adenocarcinoma - R Mirena.

Adenoma carcinoma precludes hyperbaric oxygen

- Peripheral neuropathy
- 4/1/2017 commenced STS three times per week during dialysis
- April 2017 apronectomy – chronic calciophylaxis of abdominal pannus
- Non-healing pannus wound with multiple debridements and infections. VAC dressings
- Necrotic ulcerated wounds
- Staph aureus / pseudomonas infections
- August 2018 – R foot lesions



Pathogenesis

- Uncertain
- Narrowed, calcified microvessels → chronic low-grade ischaemia
- Endothelial injury and microthrombosis → infarction
- Impaired inhibition of calcium-phosphate deposition due to deficiency of calcification inhibitors
- Carboxylated MGP is a potent inhibitor of calcification
- Carboxylation is dependent on vitamin K.



Risk Factors

- Female
- Higher BMI
- Dialysis longer than 2 years
- Lesions are similar ± ESRD
- Central lesions – abdomen, thighs – more likely if high BMI
- Peripheral lesions – digits
- Ulcerated lesions = later disease
- 6 month survival rate = 20%
- Prognosis: 1yr mortality
- ESRD = 45 – 80%
- Non- ESRD = 25 – 45%.



Elimination of Risk Factors

- Correct hypercalcemia and hyperphosphatemia
 - » cease vitamin D
 - » cease calcium based PO₄ binders
 - » decrease high calcium dialysate.
- Optimise PTH:-
 - » R calcimimetic agent
 - » parathyroidectomy.
- Increase time on dialysis
- PDX → HDX (improves control of mineral metabolism)
- Reduce local trauma – rotate s/c injection sites
- Warfarin – avoid/cease
- R Vitamin K.



Treatment:

Sodium Thiosulfate (STS)

- Antioxidant
- Vasodilator
- Blocks the ability of adipocytes to induce calcification of vascular smooth muscle cells
- ADR: volume overload, hypocalcemia, QT prolongation, hypotension, metabolic acidosis
- Optimal duration of STS therapy is unclear

Interdisciplinary Management

- Pain management
- Debridement if tolerated
- Skin graft
- Hyperbaric oxygen
- Nutritional status

Future:

- **Currently no approved therapies for calciophylaxis**
- ?re-register cinacalcet
- 2019 BEAT – Calci Study - Better Evidence and Transplantation in Calciophylaxis

An activated cell-mediated process

