CALCIPHYLAXIS

Also known as Calcific Uraemic Arteriolopathy

What is it?

Calciphylaxis is a rare and serious disorder, usually seen in individuals on treatment for severe kidney disease – typically on dialysis for end stage renal failure (ESRF).

However, calciphylaxis is not exclusive to ESRF individuals on dialysis, and can also occur in kidney transplant recipients and rarely in individuals with normal kidney function.

What causes it?

The cause of calciphylaxis is thought to be due to reduced blood flow in the small arteries (arterioles), due to abnormal calcium deposition (calcification).

Medications, chronic inflammatory conditions, and clotting disorders can act as triggers for the development of calciphylaxis

- Medications: warfarin, iron, corticosteroids, vitamin D and calcium supplements
- Chronic Inflammatory Conditions: Crohn’s Disease, connective tissue disorders, diabetes, obesity, malignancy and liver cirrhosis
- Clotting Disorders: hypercoagulable states, protein C and S deficiency, antiphospholipid syndrome

What does it look like?

The appearance of calciphylaxis depends on the time of presentation. Areas most commonly affected are the lower limbs and abdomen - especially fatty areas such as the thighs and buttocks. In the early stages, calciphylaxis usually starts as dusky purple pink, painful (tender), plaque like lesions under the skin (subcutaneous nodules), representing areas of diminished blood supply (ischaemia).

These can progress to deeper concavities with black overlying tissue (dry gangrene), representing dead tissue (eschar) from inadequate blood supply (infarction). Over time, these eschars may become infected (superinfection), leading to increased morbidity and mortality.

What other problems can occur with it?

Early changes can give a mottled appearance on the skin similar to another condition known as livedo reticularis.

Calciphylaxis can also involve the upper extremities, with lesions on the fingers occurring in some individuals.

How is it diagnosed?

The diagnosis is usually made by a dermatologist after reviewing the presentation of the individual. A skin biopsy from the lesion margin may aid or confirm diagnosis.

There are no specific blood tests for calciphylaxis. In some individuals, certain increases may be observed, although these are not always present

- High phosphate levels
- High calcium levels
- High calcium x phosphate (Ca x P) product levels
- High parathyroid hormone (PTH) levels
- An X ray may show evidence of vascular calcification within the skin.
How is it treated?

There is no consensus as to the best way to treat calciphylaxis. A multi-faceted approach to target the cause, and improve the outcomes, is recommended.

Expert wound care, monitoring for secondary infection, optimal pain relief (analgesia), and meticulous attention to minimise repetitive tissue trauma is universal. Cessation of medication predisposing to calciphylaxis is recommended, if possible.

Intravenous sodium thiosulfate may be used (this helps to remove the deposited calcium). Patients with renal failure may need some adjustments in their medication to control abnormal calcium and/or phosphate levels.

In some individuals, oxygen therapy (hyperbaric oxygen or high-flow oxygen therapy) as part of a multi-faceted approach may demonstrate benefit.

Some patients may benefit from an opinion from a surgeon for consideration of the removal of any damaged tissue.

What is the likely outcome?

The mortality rate of patients with calciphylaxis and end stage renal failure is reported to be greater than 60%-80%, often as a result of secondary infection and sepsis.