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Calciphylaxis

Calciphylaxis is a condition characterised by necrosis (cellular death) of the skin and fatty tissue. It is seen mainly in patients with end stage kidney disease

In 1981, approximately 50 cases were reported in the world literature. Today, incidence is estimated at 1 percent per year in patients undergoing dialysis. The mortality is extremely high, up to 80%, often within several months of onset. The primary cause of death is from secondary infection of the ulcers, and sepsis.

How does it occur?

The cause of calciphylaxis is not properly understood. It is associated with a condition known as secondary hyperparathyroidism. The damaged kidneys don't excrete phosphate properly, which results in a build up of phosphate in the blood, which combines with calcium. Vitamin-D levels are reduced because of the kidney failure and reduced absorption through the gut. The bones become resistant to parathyroid hormone. The parathyroid glands therefore increase in size and produce more hormone increasing the amount of calcium circulating in the blood.

Calciphylaxis can occur in those with high or normal levels of serum calcium and phosphate, with or without vitamin D replacement, in dialysed patients and less often in those who have not yet commenced dialysis or in those who have received a renal transplant. It is more common in women than in men, in obese patients compared to those of normal weight, and in patients who have been taking corticosteroids or other immunosuppressive medicines.

Small blood vessels become blocked with blood clots, which leads to the black painful necrotic areas. It is thought that the clots occur because of calcification within the walls of the blood vessels.

Clinical Features

Calciphylaxis begins as surface purple-coloured mottling of the skin then bleeding occurs within the affected area. There may be blood-filled blisters. The skin goes black in the centre of star-shaped purple lesions. The skin cells die because of lack of blood supply (dry gangrene). This causes deep and often extensive ulcers.

Calciphylaxis





Patients usually experience unbearable pain, burning and sometimes itching at the lesion sites.

Calciophylaxis most often occurs on the lower limb especially in fatty areas. Lesions on the trunk, abdomen, buttocks or thighs, appear to be more dangerous than lesions on the lower legs and feet.

Risk factors

Certain conditions in addition to renal failure are associated with accelerated calcium deposition in soft tissues:

- Obesity
- Diabetes
- Caucasian race
- Female gender
- Hypoalbuminemia

How is the diagnosis made?

A [skin biopsy](#) may be necessary to diagnose calciophylaxis as a similar appearance can be seen in other conditions such as [necrotising fasciitis](#), [cryoglobulinaemia](#), [antiphospholipid syndrome](#), coumarin necrosis and [vasculitis](#). The pathologist finds calcium within scarred and blocked blood vessels in the skin. There may also be inflammation of the fat ([panniculitis](#)).

Xrays of the affected limb may demonstrate vascular calcification within the skin; however this may also be seen in healthy patients with renal disease who are not affected by calciophylaxis.

Management

The most important initial step is to normalise the calcium - phosphate product levels, and control the hyperphosphatemia associated with renal failure. A phosphate restricted diet and dialysis with a lower dialysate calcium concentration is important initial management in the calciophylaxis associated with renal failure.

In patients with hyperparathyroidism that cannot be medically controlled, surgical removal of the parathyroid glands (parathyroidectomy) has been shown to reduce pain and promote wound healing, especially in early wound development. Parathyroidectomy should not be performed in calciophylaxis in the absence of hyperparathyroidism, as it can result in low levels of calcium in the blood and bone disease.

Some patients may also be treated with anticoagulants to reduce the tendency to form blood clots, but these are not always suitable or helpful.

Wound management is important.

- Keep wounds clean
- Surgically remove the necrotic tissue - sometimes this means a limb must be amputated
- Systemic antibiotics

Related information

On DermNet NZ:

- [Leg ulcers](#)

Other websites:

- [Calciophylaxis](#) - emedicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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