Imaging Features of Calciphylaxis

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No Disclosures

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Calciphylaxis: Background

- Calciphylaxis, also known calcific uraemic arteriolopathy, is an uncommon condition most commonly found in patients with end-stage renal disease, estimated to affect up to 1%-4% of that population and carrying a 1-year mortality rate of 45–80%

- Originally described in uremic patients it effects the small vessels causing a vasculopathy involving mural calcification with intimal fibrosis, proliferation, and thrombosis
Calciphylaxis:

Pathophysiology

- The primary lesion is the accumulating of calcium salts in the media of small arteries, causing the artery lumen to narrow.
- The luminal narrowing causes reduced arteriolar blood flow, which leads to ischemia.
- Ischemic lesions develop in the subcutaneous fat primarily involving the dermal arterioles leading to secondary subcutaneous tissue and skin infarcts and necrosis.
- On biopsy, calcium deposits are seen in media of small arteries and arterioles.
Typical clinical presentation includes ischemia and necrosis of the skin, soft tissues, and organs.

Small vessel vascular calcifications visible on imaging and are the hallmark of the disease.

Patient risk factors include: a high calcium-phosphate product, end-stage renal disease on renal replacement therapy, body mass index >30 kg/m², female gender and use of warfarin or systemic corticosteroids.
**Calciphylaxis:**

*Diagnosis*

- Classically, the diagnosis is based on the clinical presentation in combination with skin biopsy, however definitive histopathologic diagnosis is often challenging

*Imaging evaluation now plays an important complimentary role in the diagnosis of calciphylaxis!*
Objective:

We reviewed our experience with the imaging features of 11 patients with biopsy proven calciphylaxis to identify the characteristics of this uncommon entity.
Patients and Methods:

- We retrospectively reviewed all available records in our institutional electronic pathology report data base for all patients with a diagnosis of “calciphylaxis”
- A total of 43 patients were identified
- All pathology reports and imaging studies for these patients were reviewed
Patients and Methods:

- Study group inclusion criteria required:
  - Definitive pathological diagnosis of calciphylaxis or findings “strongly suggestive” of calciphylaxis and
  - Imaging studies (radiographs, CT, bone scan and/or US) of the area of the biopsy
The study group included 11 patients:
- Four men and seven women
- Average age of 52 years (range 37-77 years)

Imaging studies reviewed included:
- Radiographs (11 patients)
- CT (5 patients)
- Bone scan (2 patients)
Results:

- The dominant imaging feature was fine small vessel vascular calcification
- Calcifications were within the subcutaneous adipose most prominent deep to the skin
- These were seen well on both radiographic and CT examinations
Results:

- Skin thickening and varying amounts of associated panniculitis immediately adjacent to the skin thickening were best seen on CT.
- These findings were less well-delineated on radiographs.
- Ill-defined increased tracer accumulation was readily identified on bone scintigraphy in the areas of the calcification.
**Results:**

- Clinical photographs were available in 7 patients
- These demonstrated varying amount of skin necrosis including ulceration, involving the legs, feet, and abdominal wall
Typical Imaging Features:

Skin Thickening and Inflammation

Radiographs show extensive diabetic vascular calcification
Radiographs show extensive diabetic vascular calcification...

...and extensive small arteriole vascular calcification
Radiographs show extensive diabetic vascular calcification

...and extensive small arteriole vascular calcification

...and skin thickening with Inflammatory change
Oblique radiograph of the foot shows the necrotic ulcer over the heel to better advantage (white arrow); note small vessel calcification (black arrows).
Intraoperative image during ulcer debridement
Radiographs from a different patient show similar findings with fine small vessel calcification (arrows)
Patient developed an area of infarction
... which ulcerated
Patient developed an area of infarction
... which ulcerated
Patient developed an area of infarction which ulcerated.

Note extensive small vessel calcification (arrows).
Clinical photo shows infarction and ulceration.
Posterior image MDP bone scan with correlating CT
Increased tracer accumulation correlated to areas of vascular calcification and inflammation.
Increased tracer accumulation on the right also correlate to areas of vascular calcification (arrows) and inflammation (arrow head).
CT demonstrates extensive vascular calcifications (arrows) and edema (asterisks) with overlying ulcerations (arrowheads).
Evaluation of the ventral abdominal wall revealed ulceration and discoloration with histology showing calciphylaxis.
Calciphylaxis:

Conclusion

- Imaging findings demonstrating extensive fine small vessel vascular calcification within the subcutaneous adipose tissue, typically just deep to the skin, are characteristic of calciphylaxis.

- These findings are well seen on both radiographs and CT.
Calciphylaxis: Conclusion

- Scintigraphy will show increased tracer primarily localized to the subcutaneous tissue
- Associated inflammatory change with skin thickening is commonly seen
- Ischemia, infarction, necrosis, ulceration and infection are well recognized complications and a major cause of morbidity and mortality
References:


Calciphylaxis:

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- #Calciphylaxis