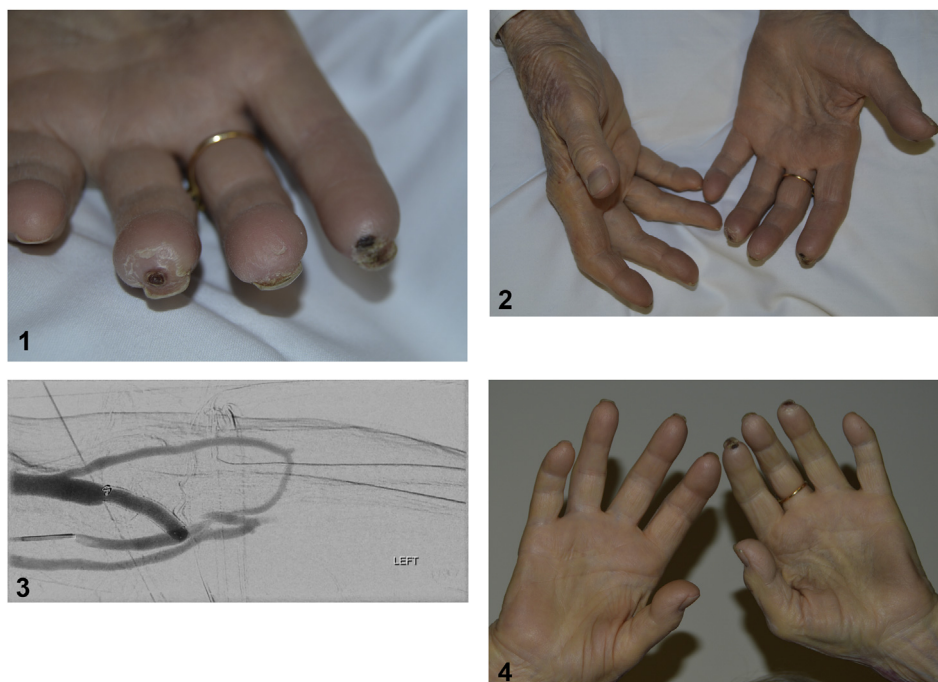


# A dialysis patient with subungual hyperkeratosis and ulceration of the fingertips



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**Key words:** arteriovenous fistula; dialysis; hyperkeratosis; ulcer development; ulceration.



An 81-year-old woman, a lifetime nonsmoker, was referred for fingernail changes concerning for onychomycosis on her left hand. She had diabetes and was on hemodialysis for nonsteroidal anti-inflammatory drug-induced nephropathy. She reported a painful left hand and fingertips for 2 months but denied weakness, cold intolerance, color changes, and numbness. The tips of her left index and ring fingers had hyperkeratotic subungual material adjacent to small crusted ulcers (Fig 1). Her right hand was uninvolved (Fig 2). Her left radial and ulnar arterial pulses were diminished; her right-sided pulses were normal. An arteriovenous hemodialysis fistula had been placed on her left antecubital fossa 4 years earlier and was functioning normally.

**Question 1: What is the most likely underlying diagnosis for the hyperkeratotic crusted ulcers of her fingertips?**

- A. Thromboangiitis obliterans (Buerger disease)
- B. Digital calciphylaxis

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Funding sources: None.

IRB approval status: Not applicable.

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JAAD Case Reports 2022;22:5-7.

2352-5126

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<https://doi.org/10.1016/j.jidcr.2021.10.040>

- C. Steal syndrome
- D. Limited systemic sclerosis
- E. Diabetic ulcer

**Answers:**

**A.** Thromboangiitis obliterans (Buerger disease) – Incorrect. This nonatherosclerotic, vaso-occlusive vasculitis is strongly associated with tobacco use and typically affects young and middle-aged men. Fingertip ulcers usually occur bilaterally.

**B.** Digital calciphylaxis – Incorrect. This is a rare occlusion syndrome of microvascular calcifications, resulting in livedo reticularis and ulcerations. The greatest risk factor is persistent hyperphosphatemia, often due to noncompliance with treatment, and warfarin use. Our patient did not have these risk factors.

**C.** Steal syndrome – Correct. Dialysis-associated steal syndrome (DASS) is an uncommon complication of long-standing arteriovenous fistulae (AVF) created for hemodialysis access. If arterial blood flow is excessively diverted through the AVF, tissues distal to the AVF are hypoperfused, causing hypoxia-induced pain, paresthesias, digital ulcers, nail changes, and even soft tissue necrosis. Onychodystrophy may be the first presenting sign of DASS.<sup>1</sup> DASS can occur acutely following AVF creation or can occur gradually as the fistula matures and dilates from intraluminal pressure.<sup>2</sup> Advanced age, female sex, and diabetes are important risk factors.<sup>3</sup> Our patient's symptoms started after she underwent a stenting procedure to correct a recently stenotic AVF.

**D.** Limited systemic sclerosis – Incorrect. In patients with limited systemic sclerosis, particularly those with calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerosis, and telangiectasias, recurrent episodes of arterial vasospasm (Raynaud phenomenon) can cause fingertip ulcers. This is a systemic disease, and its acral findings are usually bilateral and accompanied by other cutaneous findings, such as dilated nailfold capillaries.<sup>4</sup>

**E.** Diabetic ulcer – Incorrect. Diabetic ulcers arise as deep, well-defined ulcers with surrounding hyperkeratosis over pressure sites of the feet. The patient was recently diagnosed with diabetes, but her ulceration sites were atypical for diabetic ulcers. Long-standing diabetes results in decreased peripheral sensation and chronic unperceived traumas of the feet that, over time, along with peripheral

vascular disease, result in deep ulceration with high risk of secondary infection.

**Question 2: What is the gold standard for diagnosing this condition?**

- A. Duplex ultrasound with compression study
- B. Catheter angiography
- C. Computed tomography (CT) angiography
- D. Magnetic resonance imaging
- E. Skin biopsy

**Answers:**

**A.** Duplex ultrasound with compression study – Incorrect. Although the diagnosis of DASS can often be made clinically, duplex ultrasonography is a noninvasive aid to diagnosis that, when combined with a compression study (measures distal blood flow before and after external compression), can help determine interventions. Increased arterial pressure in the digits when one compresses the AVF usually indicates clinically meaningful steal syndrome, but there are no well-established hemodynamic parameters to confirm the diagnosis on ultrasound.<sup>5</sup>

**B.** Catheter angiography – Correct. Catheter angiography of the affected arteries, starting at the aortic arch, is generally accepted as the gold standard for the diagnosis of DASS. It can show a clinically significant reduction in radiographic contrast flowing from the arterial circulation across the fistula, with decreased contrast flow distally that is exacerbated by peripheral arterial disease. Catheter angiography also allows for the correction of the abnormally high flow rates and distal obstructions.<sup>2</sup> After Doppler ultrasound showed reduced distal flow rates, the patient underwent angiography, which confirmed the diagnosis of DASS (Fig 3; Video 1, available on [www.jaad.org](http://www.jaad.org)).

**C.** CT angiography – Incorrect. CT angiography may be helpful for diagnosis if catheter-based techniques are unavailable but is not yet established as a definitive diagnostic tool.

**D.** Magnetic resonance imaging – Incorrect. While magnetic resonance imaging with magnetic resonance angiography has been used for the diagnosis of DASS, it is not as well studied as invasive angiography.<sup>2</sup> Neither magnetic resonance angiography nor CT angiography allows for simultaneous intervention if indicated.

**E.** Skin biopsy — Incorrect. Skin biopsy is not the diagnostic gold standard. It is only helpful in the evaluation of digital ischemia if there is suspicion of a systemic inflammatory condition, such as endocarditis or vasculitis. Local tissue ischemia would also likely result in delayed wound healing and an increased risk of infection after biopsy.

**Question 3: What is the next best step in management?**

- A.** Surgical debridement
- B.** Referral to pain management
- C.** Referral to vascular surgery
- D.** Oral nifedipine
- E.** Compression and elevation of the affected limb

**Answers:**

**A.** Surgical debridement — Incorrect. Although the debridement of the hyperkeratotic material at the fingertips may be needed in the future, without an intervention to restore adequate blood flow, the ischemia is likely to progress, and the ulcers will recur.

**B.** Referral to pain management — Incorrect. Managing this patient's pain without correcting the underlying cause is not the optimal treatment.

**C.** Referral to vascular surgery — Correct. Several percutaneous and surgical treatment options exist for DASS, with varying degrees of evidence for each method. Catheter angiography can be performed, with angioplasty of distal atherosclerotic disease or the insertion of a coil to decrease flow rates. Surgical options including plication, banding, and clipping of the AVF to decrease shunting and restore distal flow in high-flow lesions. Recently, distal revascularization with interval ligation has become the preferred procedure for most cases of DASS.<sup>2,3</sup> Most vascular surgeons are able to perform both percutaneous and open surgical procedures

for DASS. The patient underwent banding of the fistula, with improvement in distal angiographic flow.

**D.** Oral nifedipine — Incorrect. Nifedipine is sometimes used to treat Raynaud syndrome, because it causes vasodilation and may prevent attacks. Decreasing arterial pressure in this case may make the patient's symptoms worse.<sup>6</sup>

**E.** Compression and elevation of the affected limb — Incorrect. Compression and elevation of an extremity that is not adequately perfused risks making the problem worse. Interestingly, this patient had noticeable pallor of her left hand when it was raised above her head (Fig 4).

**Abbreviations used:**

AVF: arteriovenous fistula

CT: computed tomography

DASS: dialysis-associated steal syndrome

**Conflicts of interests**

None disclosed.

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