Awareness of Scleroderma Peripheral Artery Disease Presenting With Critical Limb Ischemia in Elderly Patients

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Scleroderma is a slow progressive vascular disease with macrovascular manifestations in approximately 15% of patients.¹ An 80-year-old woman with dyslipidemia was referred for treatment of a non-healing ulcer in the right first and fifth toes (**Figure A**). Right foot skin perfusion pressure was 17/16 mmHg (dorsum/plantar), suggesting unlikely wound healing. Diagnostic angiography showed severe infrapopliteal artery disease with a type 1C variant (**Figure B**). Given the isolated infrapopliteal artery disease without diabetes mellitus and renal failure, non-atherosclerotic vasculopathy was highly suspected. Following a rheumatology consultation, the diagnosis of limited scleroderma was made with the evidence of anti-centromere antibody 169 index (normal high >16), skin thickening and Raynaud's phenomenon. Medical treatment including antiplatelet agent and endothelin receptor antagonist failed to improve the clinical condition. Therefore, although surgical or endovascular recanalization might be controversial in the setting of vasculitis,^{2,3} endovascular recanalization was carried out. The occlusive anterior tibial and dorsalis pedis arteries were successfully recanalized to establish one straight-line flow, whereas the occlusive posterior tibial and plantar arteries were not (**Figure C,D**). The skin perfusion pressure increased to 32/20 mmHg on the following day. The wound healed completely in 3 months with 3 clinically driven reinterventions and a scheduled minor amputation (**Figure E**). Histopathology of the

amputated toe indicated concentric intimal hyperplasia leading to narrowing of the lumen in the digital artery, suggesting an underlying vasculitis (**Figure F**). Endothelin receptor antagonist as well as antiplatelet agent were given to prevent the recurrence of toe ulcer. During the last 6 months, no recurrence of the wound was observed.

This emphasizes the need for an increasing awareness of scleroderma-related peripheral artery disease (PAD) in elderly patients. Endovascular therapy might be a potential treatment option even in the setting of non-atherosclerotic PAD.

Disclosures

The authors declare no conflicts of interest.

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Figure. Scleroderma-related isolated infrapopliteal artery disease presenting with ischemic toe ulcer in an 80-year-old woman. (**A**) Non-healing ischemic ulcer on the right great and little toes. (**B**) Diagnostic angiography showed severe infrapopliteal artery disease with a type 1C variant consisting of an extensive occlusion from the ostium of the right anterior tibial artery to the dorsalis pedis artery (arrow), diffuse stenosis in the posterior tibial artery, and occlusion of the plantar artery. (**C,D**) Following balloon angioplasty of the anterior tibial and dorsalis pedis arteries, final angiography demonstrated an excellent straight-line flow to the foot and the establishment of pedal arch. The lesser visualization of the branches might be due to underlying small vessel disease or vasospasm related to vasculitis. (**E**) Complete wound healing after endovascular intervention and scheduled minor amputation. (**F**) Histopathology of the amputated toe showing concentric intimal hyperplasia leading to luminal stenosis or occlusion in the digital artery, suggesting an underlying vasculitis.