Painful blisters on fingertips and toes

Our patient had visited the emergency department for painful blisters on her fingertips and toes. A follow-up visit to our clinic unearthed the cause.

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A 52-YEAR-OLD WOMAN presented to the emergency department (ED) with a 4-month history of recurrent painful blisters on her fingertips and the tips of her toes (**FIGURE 1**), arthralgias, painful discoloration of her distal toes and fingers when exposed to cold, and painful nodules on her forearms. She was started on prednisone and was sent to our clinic for follow-up.

At her initial visit to our office, she was continued on prednisone and referred to Rheumatology and Interventional Cardiology, where a work-up for rheumatoid arthritis, systemic lupus erythematosus, and other vasculitides was negative. The patient had normal arterial pressures and a normal echocardiogram. An angiogram revealed segmental occlusions of the distal vessels in her arms and legs. The patient denied chest pain, syncope, dyspnea on exertion, or fever. She reported a >30 pack-year history of cigarette smoking.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE 1
Painful blisters on fingertips

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Diagnosis: Thromboangiitis obliterans

Thromboangiitis obliterans (TAO), or Buerger's disease, is a rare nonatherosclerotic disease that affects the medium and small arteries. The disease has a male predominance, primarily occurs in those younger than 45 years of age, and is most common in people from the Middle and Far East.[1] Its distinctive features include ulcerations of the distal extremities and symptoms of claudication and pain at rest. More than 40% of affected patients develop Raynaud's phenomenon.[1] Superficial thrombophlebitis in the form of painful nodules has also been described.[2]

The etiology of TAO is likely due to disordered inflammation of endothelial cells, which has a strong association with smoking.[3] The exact pathogenesis is unknown, but genetics and autoimmunity are suspected contributing factors.

The diagnosis is based on exclusion of other causes

The differential diagnosis includes diabetic angiopathy, embolic disease, atherosclerosis, hypercoagulability/thrombophilia, vasculitis or connective tissue diseases, and drug-associated (eg, cocaine) vasculitis.[4]

The diagnosis of TAO is based on the exclusion of other causes, although several diagnostic criteria have been proposed, including:

- age <45 years
- current or recent history of tobacco use
- distal extremity involvement (ulcers, claudication, or pain at rest)
- exclusion of diabetes, peripheral artery disease, thrombophilia, or embolic disease
- typical arteriographic findings on imaging, including distal small to medium vessel involvement, segmental occlusions, and "corkscrew-shaped" collaterals.[1,2,5,6]

Lab tests. There are no specific laboratory markers for TAO. The initial evaluation should include an erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), complete metabolic panel (CMP), and urinalysis (UA). Tests to exclude other autoimmune diseases include rheumatoid factor, antinuclear antibody, anticentromere antibody and Scl-70 to exclude CREST syndrome and scleroderma, antiphospholipid antibodies to exclude disorders of hypercoagulability, and drug testing and history-taking to evaluate for drug-related (eg, cocaine) etiologies. Further studies should be performed based on clinical suspicion.

Imaging. Patients with suspected TAO should undergo an arteriogram of the affected extremities and large arteries. Other imaging modalities include computed tomographic angiography and magnetic resonance angiography. Biopsy is rarely indicated, unless there are atypical findings, such as large artery involvement or arterial nodules. Interestingly, a positive Allen test in a young smoker can be highly suggestive of TAO.[1] (For a demonstration of the Allen test, see https://www.youtube.com/watch?v=D1tJO0RW9UM.)

Our patient tested negative for rheumatoid arthritis, CREST, and scleroderma and had a normal UA and CMP. She did have a slightly elevated anticardiolipin antibody test, but a negative lupus anticoagulant test, the significance of which is uncertain. Her CRP and ESR were elevated.

Complete smoking cessation is essential for treatment

Several treatments have been proposed, including prostanoids and surgery (surgical revascularization or endovascular therapy).[1,4]

FIGURE 2
2 months after smoking cessation

The patient's pain and ulcerations had almost completely resolved 2 months after she quit smoking.

In severe cases, amputation may be required to remove the affected extremity. However, the most important and most effective treatment for TAO is smoking cessation.[1] Of note, several case reports have found that replacing smoking with other nicotine-containing products (eg, chewing tobacco) may not prevent limb loss.[7-9]

Our patient was tapered off prednisone and was continued on amlodipine 5 mg/d for vasospasm. She was started on varenicline 0.5 mg/d, which was increased to twice daily by Day 4 to aid with smoking cessation. Two months later, the patient's pain and ulcerations had almost completely resolved (**FIGURE 2**). She experienced occasional relapses with smoking, during which her ulcerations and Raynaud's would return. This case reinforces the age-old aphorism of "no tobacco, no Buerger's disease."[4] **JFP**

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References

1. Olin JW. Thromboangiitis obliterans (Buerger's disease). *N Engl J Med*. 2000;343:864-869.
2. Piazza G, Creager MA. Thromboangiitis obliterans. *Circulation*. 2010;121:1858-1861.
3. Azizi M, Boutouyrie P, Bura-Rivière A, et al. Thromboangiitis obliterans and endothelial function. *Eur J Clin Invest*. 2010;40:518-526.
4. Klein-Weigel PF, Richter JG. Thromboangiitis obliterans (Buerger's disease). *Vasa*. 2014;43:337-346.
5. Papa MZ, Rabi I, Adar R. A point scoring system for the clinical diagnosis of Buerger's disease. *Eur J Vasc Endovasc Surg*. 1996;11:335-339.
6. Mills JL, Porter JM. Buerger's disease: a review and update. *Semin Vasc Surg*. 1993;6:14-23.
7. Lie JT. Thromboangiitis obliterans (Buerger's disease) and smokeless tobacco. *Arthritis Rheum*. 1988;31:812-813.
8. O'Dell JR, Linder J, Markin RS, et al. Thromboangiitis obliterans (Buerger's disease) and smokeless tobacco. *Arthritis Rheum*. 1987;30:1054-1056.
9. Lawrence PF, Lund OI, Jimenez JC, et al. Substitution of smokeless tobacco for cigarettes in Buerger's disease does not prevent limb loss. *J Vasc Surg*. 2008;48:210-212.

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