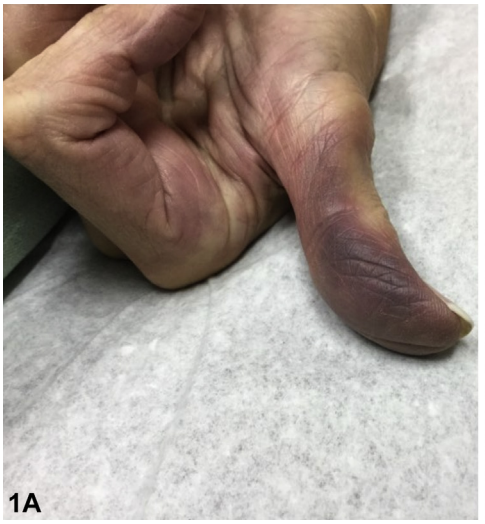
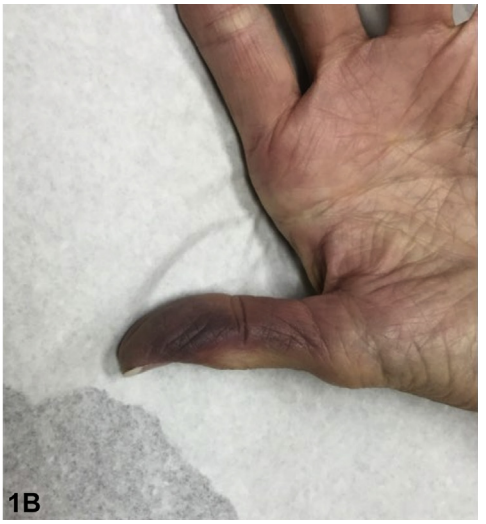
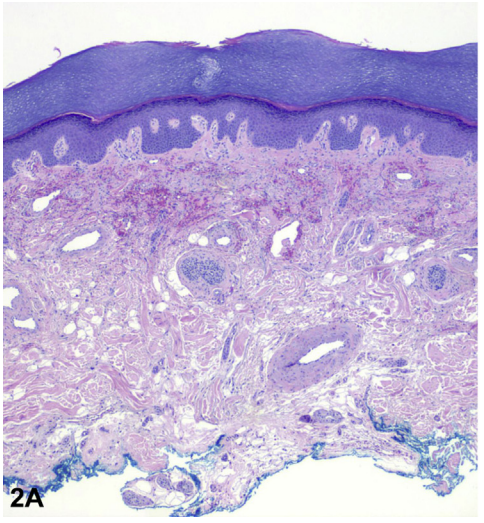
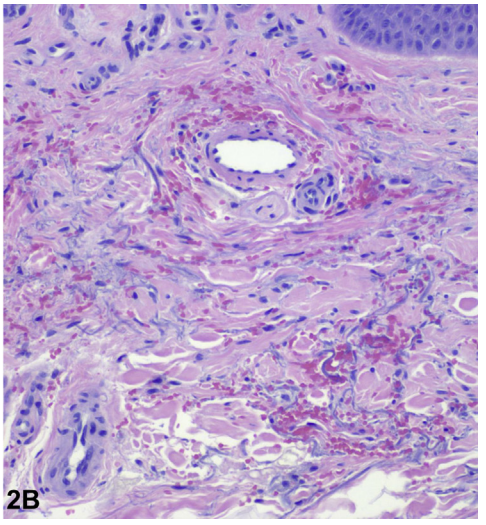
The case of a painful blue thumb

Sepideh Ashrafzadeh, BS,[a] Yun Xue, MD,[a] Thomas D. Horn, MD,[a,b] and John B. Korman, MD[c]
Boston, Massachusetts and Greer, South Carolina









From the Department of Dermatology[a] and Department of Pathology,[b] Massachusetts General Hospital, Boston; and Upstate Dermatology, Greer.[c]
Funding sources: None.
Conflicts of interest: None disclosed.
Correspondence to: John B. Korman, MD, Upstate Dermatology, 420 The Parkway, Ste M, Greer, SC 29650. E-mail: johnkormanmd@hotmail.com.

JAAD Case Reports 2020;6:1053-5.
2352-5126

https://doi.org/10.1016/j.jdcr.2020.06.045

BRIEF HISTORY

A 65-year-old woman presented for evaluation of a tender, purple discoloration of her left thumb and index finger that had recurred monthly for 3 years (Fig 1). Her symptoms started as an intense burning sensation followed by mild swelling and purple discoloration that spontaneously resolve after 3 to 7 days. She had no systemic symptoms or triggers such as trauma, cold temperatures, or medications. She received metoprolol for supraventricular tachycardia and had Raynaud syndrome, which presented differently. Physical examination revealed a purpuric patch on the palmar aspect of the left thumb, extending onto the dorsal distal aspect of the phalanx. A punch biopsy was performed.

Question 1: What is the most likely diagnosis?

A. Raynaud syndrome

B. Polyarteritis nodosa

C. Arterial embolism

D. Achenbach syndrome

E. Traumatic hematoma

Answers:

A. Raynaud syndrome—Incorrect. Raynaud syndrome is the abnormal vasoconstriction of digital arteries and arterioles because of cold temperatures, emotional stress, or systemic disease. Although it may cause pain, it usually lasts for 15 to 20 minutes and is characterized by multiple discolorations (eg, white, blue, red).

B. Polyarteritis nodosa—Incorrect. Several vasculitides (eg, Takayasu arteritis, cutaneous leukocytoclastic vasculitis, polyarteritis nodosa, microscopic polyangiitis) may result in digital purpura. However, these vasculitides are often associated with other systemic findings and do not follow a benign cyclical pattern.[1]

C. Arterial embolism—Incorrect. Although an arterial embolus causes acute pain and discoloration, patients typically have subsequent tissue necrosis, rather than spontaneous resolution, without intervention.

D. Achenbach syndrome—Correct. This is a case of Achenbach syndrome, a rare, self-limiting condition of unknown etiology that predominantly affects middle-aged women.[2,3] The syndrome is characterized by recurrent paroxysmal hematoma in the fingers, often in the left hand.[2,3] Clinically, patients present with a sudden burning pain and cyanosis in 1 or more digits (usually the index or middle finger, and less commonly the thumb) that may be associated with swelling, pruritus, coldness, and restricted digit movement, followed by spontaneous resolution without treatment.[2-4] In the absence of concerning history or alarm symptoms and signs (eg, fever, hypertension, nonpalpable pulses, cutaneous necrosis or ulceration), the diagnosis is established clinically and does not require further testing or procedures.[4]

E. Traumatic hematoma—Incorrect. The patient denied history of repeated trauma or conditions that increase vascular fragility such as Ehlers-Danlos syndrome.

Question 2: What would you expect to find in the diagnostic evaluation of this case?

A. Elevated prothrombin time

B. Normal complete blood cell count

C. Atrial fibrillation on electrocardiogram

D. Elevated C-reactive protein and erythrocyte sedimentation rate

E. Venous thrombosis on left-sided upper extremity ultrasonography

Answers:

A. Elevated prothrombin time—Incorrect. An elevated prothrombin time would be expected if the patient had a history of anticoagulant use, liver disease, vitamin K deficiency, or coagulation factor deficiency. Patients with an elevated prothrombin time may report severe and frequent bleeding and present with hemarthrosis and extensive ecchymoses, whereas this patient reported only localized symptoms in her thumb.

B. Normal complete blood cell count—Correct. Achenbach syndrome is associated with normal complete blood cell counts, as well as normal coagulation parameters and inflammatory markers.[3,4]

C. Atrial fibrillation on electrocardiogram—Incorrect. This finding would be expected if the patient's symptoms were secondary to arterial emboli. However, arterial occlusion would result in irreversible tissue necrosis and require intervention, neither of which was true in this case because this patient's symptoms return to baseline between episodes.

D. Elevated erythrocyte sedimentation rate—Incorrect. We would expect this finding if the underlying etiology were a vasculitis or other rheumatologic disease contributing to secondary Raynaud syndrome. However, given the absence of systemic and histopathologic findings, neither of these diagnoses is likely.

E. Venous thrombosis on left-sided upper extremity ultrasonography—Incorrect. Although a venous thrombus can cause swelling, pain, and color changes to the finger, these symptoms would not recur or self-resolve monthly for several years, as in this case. Also, the patient did not have risk factors such as trauma, surgery, or recent immobility to place her at risk for venous thrombosis.

Question 3: Which of the following histologic findings do you see in this patient's biopsy in Fig 2?

A. Dermal hemorrhage with intact vessel walls

B. Dermal vessels with neutrophilic inflammatory infiltrate, fibrin deposition, and red blood cell (RBC) extravasation

C. Occlusive, highly cellular and inflammatory thrombus with sparing of the vessel wall

D. Multiple arteriovenous anastomoses with connective tissue edema and fibrosis

E. Normal tissue and vessel architecture

Answers:

A. Dermal hemorrhage with intact vessel walls—Correct. The histologic findings associated with Achenbach syndrome are RBCs in the dermis and possibly below the dermis, without evidence of vascular damage.[4]

B. Dermal vessels with neutrophilic inflammatory infiltrate, fibrin deposition, and RBC extravasation—Incorrect. This finding would be expected with cutaneous leukocytoclastic vasculitis, but Fig 2 shows neither inflammatory cells nor vessel wall damage.

C. Occlusive, highly cellular and inflammatory thrombus with sparing of the vessel wall—Incorrect. This histologic description is consistent with the acute phase of Buerger disease, which is also known as thromboangiitis obliterans. Buerger disease is characterized by inflammatory thrombi in small- and medium-sized arteries and veins, an intact internal elastic lamina, and possible disruption of the external elastic lamina. Although the vessel walls in Fig 2 are intact, the vessel lumen is not obstructed with a thrombus.

D. Multiple arteriovenous anastomoses with connective tissue edema and fibrosis—Incorrect. This is a nonspecific finding that may be associated with acrocyanosis, a condition associated with symmetric, persistent, blue discoloration in the distal aspect of the fingers and toes secondary to diminished oxyhemoglobin supply.[5] There are no arteriovenous anastomoses or fibrosis apparent in Fig 2.

E. Normal tissue and vessel architecture—Incorrect. This description would be an expected histologic finding for Raynaud syndrome. The histopathologic findings of Achenbach syndrome reveal dermal RBCs.

Abbreviation used:

RBC: red blood cell

REFERENCES

1. Carlson JA. The histological assessment of cutaneous vasculitis. *Histopathology*. 2010;56(1):3-23.
2. Ahmed Z, Elmallah A, Elnagar M, Dowdall J, Barry M, Sheehan SJ. Painful blue finger-Achenbach's syndrome: two case reports. *EJVES Short Rep*. 2018;40:1-2.
3. Ada F, Kasimzade F. Analysis of 24 patients with Achenbach's syndrome. *World J Clin Cases*. 2019;7(10):1103-1110.
4. Takeuchi H, Uchida HA, Okuyama Y, Wada J. Acute idiopathic blue fingers: a young man with Achenbach's syndrome. *BMJ Case Rep*. 2016;2016. https://doi.org/10.1136/bcr-2016-214491.
5. Kurklinsky AK, Miller VM, Rooke TW. Acrocyanosis: the Flying Dutchman. *Vasc Med*. 2011;16(4):288-301.