

The Medicine Forum

Volume 24

Article 6

2023

Achenbach Syndrome: A Classic Presentation of a Not-So-Common Condition

Brandon Pecchia, MD, MS Thomas Jefferson University, brandon.pecchia@jefferson.edu

Follow this and additional works at: https://jdc.jefferson.edu/tmf

Part of the Internal Medicine Commons
<u>Let us know how access to this document benefits you</u>

Recommended Citation

Pecchia, MD, MS, Brandon (2023) "Achenbach Syndrome: A Classic Presentation of a Not-So-Common Condition," *The Medicine Forum*: Vol. 24, Article 6. DOI: https://doi.org/10.29046/TMF.024.1.005 Available at: https://jdc.jefferson.edu/tmf/vol24/iss1/6

This Article is brought to you for free and open access by the Jefferson Digital Commons. The Jefferson Digital Commons is a service of Thomas Jefferson University's Center for Teaching and Learning (CTL). The Commons is a showcase for Jefferson books and journals, peer-reviewed scholarly publications, unique historical collections from the University archives, and teaching tools. The Jefferson Digital Commons allows researchers and interested readers anywhere in the world to learn about and keep up to date with Jefferson scholarship. This article has been accepted for inclusion in The Medicine Forum by an authorized administrator of the Jefferson Digital Commons. For more information, please contact: JeffersonDigitalCommons@jefferson.edu.

Achenbach Syndrome: A Classic Presentation of a Not-So-Common Condition

Brandon Pecchia, MD, MS¹

1. Department of Medicine, Thomas Jefferson University Hospital, Philadelphia, PA

INTRODUCTION

Also called "acute idiopathic blue finger" or "paroxysmal finger hematoma," Achenbach syndrome is a benign collection of findings that is often mistaken for more serious conditions. Characteristically presenting with discrete unexplained bruising or discoloration of the volar aspect of one or two fingers, it is a diagnosis that physicians should include on their differential given its innocuous course and excellent outcomes, without need for invasive testing or intervention. The purpose of this case report therefore is to increase awareness of this rare condition, especially among emergency department physicians and internists, in order to minimize the incidence of unnecessary testing, procedures, and psychological burden.

CASE PRESENTATION

Subjective:

A 37-year-old female with a past medical history of hypertension, gastroesophageal reflux disease, and anxiety, presented to the Thomas Jefferson University Hospital emergency department with a few hours of mild throbbing pain, swelling, and darkened discoloration of her right second and fifth digit. She did notice that there was brief whitening of the digits preceding the darkening discoloration. She described the sensation as an uncomfortable feeling "like a rubber band" on her wrist. She also endorsed right hand numbness that extended up the right forearm. She could not identify any specific trigger, including cold exposure or trauma. There was no other bruising or bleeding she noticed. The patient also reported that she had one very similar experience three years prior, involving the first and second digit on the right hand. At that time, she presented to another hospital, where she was evaluated by vascular surgery. She noted that she underwent catheter-directed thrombolysis, was given prednisone and gabapentin, and discharged on aspirin and clopidogrel for a few months.

Upon review of systems, she denied any other paresthesias, subjective fevers or chills, chest pain, palpitations, shortness of breath, or gastrointestinal symptoms. She denied any nailbed changes (including splinter hemorrhages) or fingertip discoloration that she had noticed. She denied any known history of heart disease. She had a history of tobacco use (around 7 pack



Figure 1: Volar and dorsal aspects of patient's right hand.

years) but had quit three years prior. She endorsed intermittent social alcohol use, but denied ever using illicit drugs, including intravenous drug use. Besides an unknown type of stroke in her grandmother at an advanced age, she denied any family history of any clotting or bleeding disorders, spontaneous abortions or miscarriages, or myocardial infarctions. She had a family history of prostate cancer in her father, and breast cancer in her maternal grandmother. She had not taken any oral contraceptives in six months and had no other hormonal contraceptive use. She was taking nifedipine for her hypertension. She had no pertinent past surgical history.

Objective:

On presentation, she was slightly tachycardic to 103, and hypertensive to 149/81. Her physical exam was notable for purple, non-blanching discoloration of the palmar surface of the second and fifth digits on the right hand, well demarcated between the proximal and distal interphalangeal joints, with distal sparing (figure 1). There was minimal swelling and tenderness in this region. Her skin was warm and dry, and sensation was intact throughout all dermatomes of her right upper extremity. She had complete ability to flex and extend all fingers. There was no distal finger tenderness or ulceration. Kanavel's signs for infectious tenosynovitis (preferred flexed positioning, fusiform digital swelling, tendon sheath tenderness, and pain with passive extension) were negative. Pulses were palpable and capillary refill was brisk. The remainder of her physical exam was within normal limits. Her ANA was negative, and CBC, PT/INR, PTT, and ESR were all within normal limits. Xray of the hand was performed (figure 2), followed by arterial and venous ultrasound of the right upper extremity (figure 3), all of which were normal.



Figure 2: X-ray of right hand: No acute fracture or joint malalignment. Negative ulnar variance.



Figure 3: Sample image of patient's right ulnar artery. Ultrasound of the right upper extremity arteries and veins (with doppler) revealed absence of deep vein thrombosis; with compressible veins, normal color filling, and normal spectral doppler waveforms. The radial and ulnar arteries and visualized digital arteries (including the second- and fifth-digit arteries) were within normal limits.

DISCUSSION

As with many medical conditions, there is a common or "textbook" presentation of Achenbach syndrome, and this patient exhibited many of those classic features. This includes blue-purple discoloration most often affecting the right index finger in middle-aged females; sparing of the distal phalanx and nailbed, as well as dorsum of the hand; discontinuous finger involvement; warm and well-perfused digits; absence of any obvious trigger; and most importantly, self-resolution in three to six days requiring no intervention and with no residual sequelae. Prodromal symptoms may be present, including pain and paresthesias. First described by Dr. Walter Achenbach in 1958 – then called "paroxysmal hand hematoma" or "finger apoplexy" - the etiology is not clearly understood, and the only pathologic findings are capillary rupture. No biopsy or intravascular intervention is needed. With fewer than 100 cases reported in the medical literature, the diagnosis is clinical, based on the common features, as routine testing (including immunologic) is usually unremarkable.

For this patient, hand surgery was consulted, who fortunately recognized the signs and symptoms fitting the description of Achenbach syndrome. Although initially given oxycodone, the patient was soon advised of the benign nature of the condition, and was discharged home with reassurance and acetaminophen for pain relief. She was encouraged to return to the emergency department if her symptoms worsened or failed to resolve; and otherwise advised to follow up with her primary care provider. No subsequent medical records documented persistence or further intervention that she required. While it is indeed a benign condition, it is necessary nonetheless to rule out more critical diagnoses that require further work up and treatment, including acute limb ischemia, fracture, thromboembolic phenomena, cellulitis, septic thrombi, hypothenar hammer syndrome, and acrocyanosis, among others. In addition to good history taking and physical exam, the patient should generally receive radiographs, an arterial and venous ultrasound of the extremity with doppler, and basic lab work to help rule out infection, signs of systemic inflammation, or immunologic disease. The patient in this case did not have especially convincing familial or personal concerns for thrombosis or bleeding, nor signs or symptoms of systemic illness or infection. Achenbach syndrome is often misdiagnosed as Raynaud's, as they both include finger discoloration with a higher propensity in females, and can both present with a preceding pale phase. Raynaud's however is more often chronic and episodic, frequently in response to a temperature trigger that improves with rewarming, and can be related to immunologic disease. For the patient in the case, there was no cold exposure or stress trigger to bring about this issue, and the specific geographic pattern of her presentation was less fitting. Thromboangiitis obliterans usually occurs with tobacco exposure, resulting in ulceration and gangrene of the digits. Although the patient in this case was a prior smoker, this diagnosis is much less common in females, and she had no ulceration or arterial occlusions. Hypothenar hammer syndrome is often seen in males and is the result of repetitive palmar trauma (denied by this patient), such as with occupational overuse. Systemic vasculitis may be more difficult to differentiate, but often has intense pain and necrosis, and the patients typically have lab changes not present in this case.

While it has been known to recur in some people, as with this patient, the prognosis for Achenbach syndrome is excellent. Given this patient's prior presentation at another hospital and subsequent involved workup, possibly unnecessary intervention (although outside records were unavailable), and not risk-free pharmacotherapy, the importance of recognizing this syndromic presentation and including it in the differential is key. It is not routinely included in medical school curricula and, as seen in this case, is rare enough that emergency department providers and even vascular surgeons may not recognize it. As more cases are documented, the informed internist and emergency medicine provider should be aware of this condition, know basic testing to perform to rule out more severe disease, and be able to provide assurance to their patients of its benign nature without the need for extensive testing or invasive procedures.

REFERENCES

- 1. Godoy A, Tabares AH. Achenbach syndrome (paroxysmal finger hematoma). Vasc Med. 2019 Aug;24(4):361-366.
- Kordzadeh A, Caine PL, Jonas A, Rhodes KM, Panayiotopolous YP. Is Achenbach's syndrome a surgical emergency? A systematic review. Eur J Trauma Emerg Surg. 2016 Aug;42(4):439-443.
- Moss C, Cohen SN. Achenbach syndrome: no need for skin biopsy. Clin Exp Dermatol. 2022 Sep;47(9):1730.
- Notomi K, Harada T. Achenbach syndrome. CMAJ. 2019 May 27;191(21):E584.
- Sigha B, Josselin L, Gatfosse M, Fardet L. Hématomes digitaux spontanés paroxystiques (syndrome d'Achenbach) [Paroxysmal finger haematoma (Achenbach syndrome)]. Ann Dermatol Venereol. 2016 Feb;143(2):130-3.
- Trommeter RA, Freeman CL, Shah KS, McKinzie JP, Smith AT. Hypothenar Hammer Syndrome. J Emerg Med. 2019 Jan;56(1):105-106.