BRIEF ARTICLE

A Rare Presentation of Self-Resolving Purple Fingers

Chelsea Taylor Schwartz, DO[1], Raj Watson, BS[2], David Cleaver, DO, FAAD, FAOCD[3]

[1] Still OPTI/ Northeast Regional Medical Center
[2] Kirksville College of Osteopathic Medicine
[3] Cleaver Dermatology

ABSTRACT

Achenbach syndrome is a rare, benign, self-limiting, syndrome of unknown etiology. Here, we report a 66-year-old female who presented to the dermatology clinic with waxing and waning non-palpable purpura for the last year on the volar aspect of her hands bilaterally. The patient admitted to tingling prior to the onset of the purpura and mild tenderness following presentation. The purpura would spontaneously resolve without discoloration or scarring after 2-7 days. The patient stated these episodes can occur anywhere from a few times a month to every 1-2 months. Evaluation and work-up performed by her primary care physician and rheumatologist did not reveal an explanation for her symptoms. Two punch biopsies were performed and revealed a perivascular lymphocytic infiltrate with extravasted erythrocytes and pigmented macrophages consistent with a pigmented purpuric dermatosis. Immunofluorescence was negative. The diagnosis of Achenbach syndrome was made by diagnosis of exclusion. Reassurance was provided to the patient and she was given DerMend Bruise formula cream, which contains retinol, glycolic acid, arnica oil, ceramides, niacinamide, and phytonadione, to monitor for improvement of her symptoms. It is important for healthcare providers to be aware of the benign nature of this syndrome to be able to provide reassurance to patients and avoid an unnecessary, more invasive evaluation.

INTRODUCTION

Achenbach syndrome, also known as paroxysmal finger hematoma, is a rare, benign, self-limiting condition of unknown etiology. Patients typically present with episodic pain, swelling, and paresthesias in one or more digits accompanied by a subsequent hematoma localized to the palmar aspect of the digits. The hematoma spontaneously resolves within 2-14 days without going through the typical stages of ecchymosis. Risk factors such as trauma, bleeding disorder, or rheumatologic disease are not known to be associated with the etiology of Achenbach syndrome. It is thought that rupture of superficial veins and resultant hemorrhage can cause the signs and symptoms of the disease. We report a case of a 66-year-old female diagnosed with Achenbach syndrome after excluding other possible diagnoses.

CASE REPORT

A 66-year-old Caucasian female presented to the dermatology clinic with waxing and waning non-palpable, purpuric, patches on the volar aspect of her hands bilaterally (**Figure 1**). She stated the purpuric patches

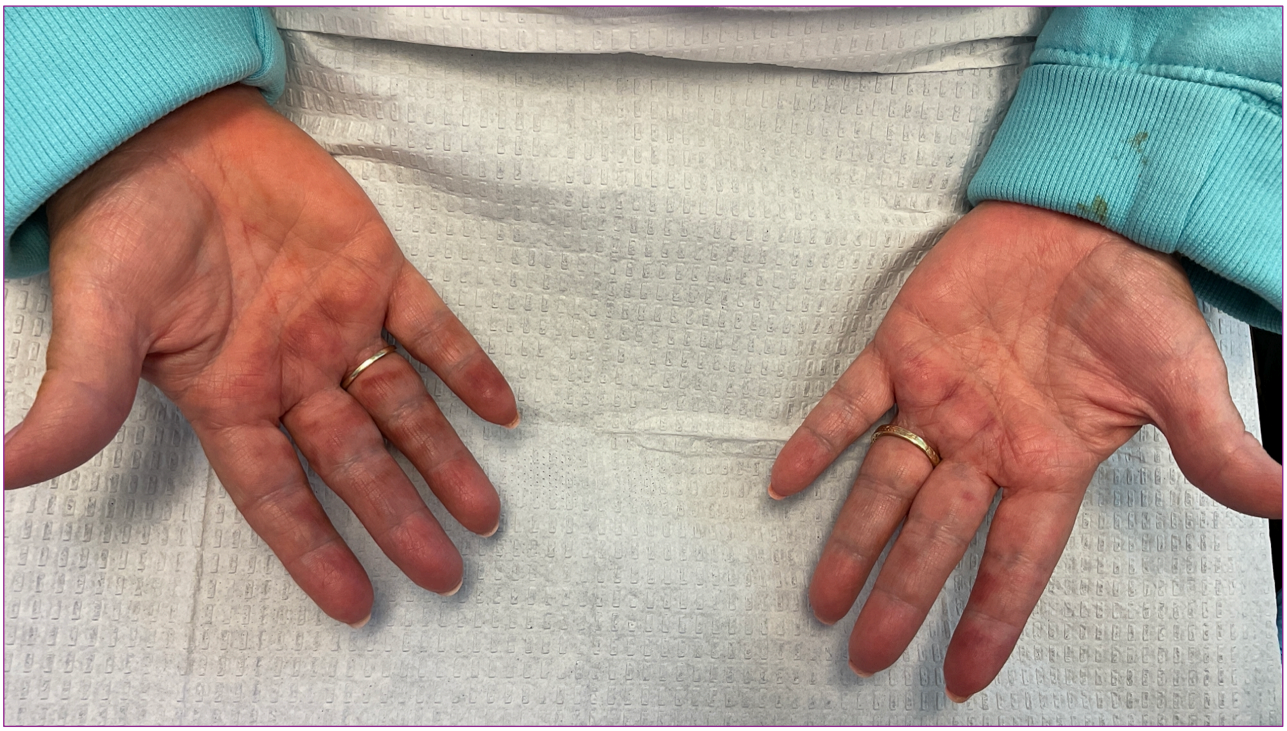

Figure 1. Non-palpable, purpuric, patches on the volar aspect of multiple digits bilaterally.

presented 1.5 years prior. She experienced tingling prior to the onset of the lesions and mild tenderness when the purpura was present. She denied any trauma to her hands. The episodes would occur from multiple times a month to monthly and spontaneously resolve within 2-7 days. No residual pigmentation was present after the lesions resolved.

The patient was evaluated by rheumatology prior to being evaluated by dermatology. Rheumatology ordered a complete blood count, complete metabolic panel, and coagulation studies which were unremarkable. Additional labs revealed a normal erythrocyte sedimentation rate (ESR) 27 and C-reactive protein (CRP) <3.0. Antinuclear antibody (ANA) , anti-double stranded DNA (dsDNA), anti-Jo 1, anti-Sjogren's syndrome A (SSA/Ro), anti-Sjogren's syndrome B (SSB/La), anti-ribonucleoprotein (RNP), anti-scleroderma 70 (Scl-70), anti-smith antibody (Sm), anti-myeloperoxidase, and anti-proteinase 3 (PR3) were all negative. Anti-cardiolipin antibodies were also negative and complement studies were normal. Rheumatology did not find any evidence of vasculitis or connective tissue disease. A punch biopsy of the lesion was performed in the dermatology clinic. Hematoxylin and eosin revealed a perivascular lymphocytic infiltrate, extravasated erythrocytes, and pigmented macrophages consistent with a pigmented purpuric dermatosis (**Figures 2 and 3**). Immunofluorescent studies were negative.

The patient was diagnosed with Achenbach syndrome by diagnosis of exclusion. The patient was reassured of the benign nature of this syndrome and that no further workup was required. She was given DerMend bruise formula cream containing retinol, glycolic acid, arnica oil, ceramides, niacinamide, and phytonadione to use when her symptoms recurred.

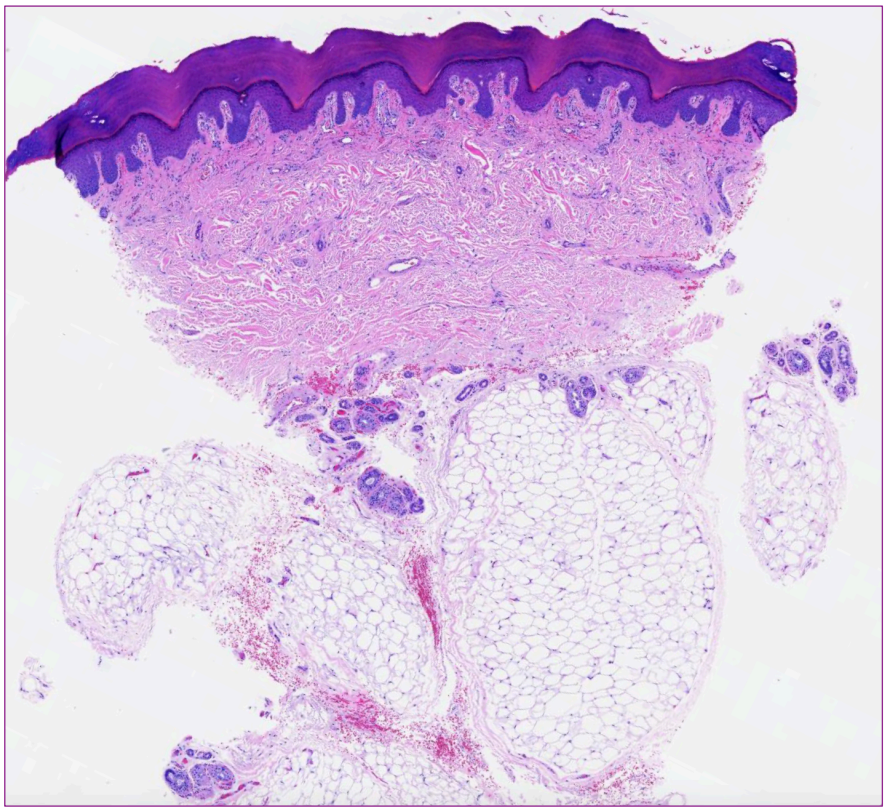

Figure 2. Punch biopsy of lesion stained with hematoxylin & eosin. Dermatopathology revealed a perivascular lymphocytic infiltrate, extravasated erythrocytes, and pigmented macrophages consistent with a pigmented purpuric dermatosis.

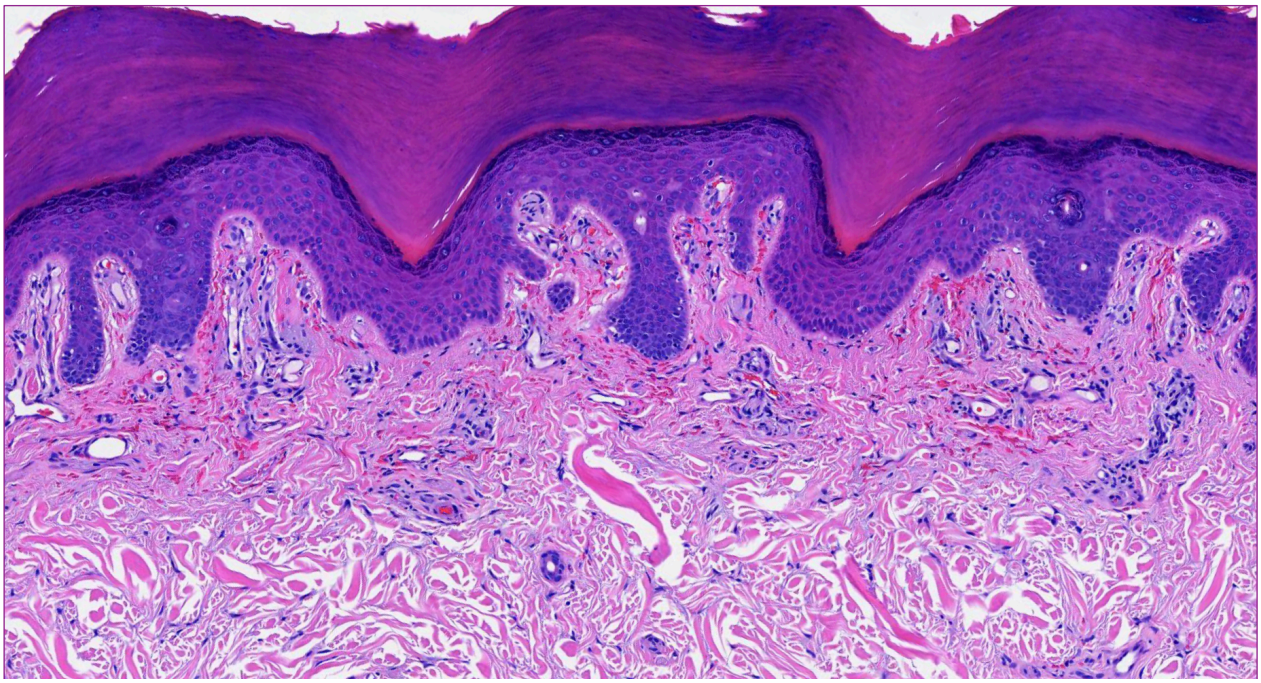

Figure 3. Higher power view of punch biopsy from acral skin revealing a perivascular lymphocytic infiltrate and extravasated red blood cells.

DISCUSSION

Achenbach syndrome, first described in the 1950's by German physician Walter Achenbach, is a rare, benign condition characterized by episodes of pain, swelling, and paresthesia followed by purpuric discoloration of the palmar aspect of the digits.[1] Presenting more often in women around their 5th decade of life, the discoloration of Achenbach syndrome typically resolves without treatment between 2 and 14 days.[2] The most common location of presentation are the index and middle fingers, although, it has been reported that discoloration can impact the thumb, dorsal surface of the hand, and even the feet and toes.[3] The etiology of the disease is not clear, but it is thought to be caused by fragility of superficial veins with resultant rupture and hemorrhage.[4] Risk factors such as trauma, drug use, bleeding disorder, or rheumatologic disease are not known to be associated with Achenbach syndrome. However, history of acrocyanosis, gastrointestinal disorders, migraines, and gallbladder disease have been reported associations in the literature.[5,6] Additionally, genes responsible for blood coagulation and inflammation, namely the F3, F2, and CRP genes, have been weakly linked to familial Achenbach syndrome.[7]

Diagnostic workup of a case of Achenbach syndrome will reveal normal complete blood count, coagulation markers, and inflammatory markers.[8] Allen's test, arterial-brachial index, pulse oximetry, a Doppler ultrasound, and an echocardiogram will all be normal. Included in the differential diagnosis will be trauma, acute limb ischemia, thrombosis, Raynaud's syndrome, auto-erythrocyte sensitization syndrome, factitious dermatitis, vasculitis, acrocyanosis, Gardner-Diamond syndrome, erythromelalgia, pernio, and drug-induced hematoma. Histopathology slides denote dermal hemorrhage, extravasated erythrocytes, and no evidence of vascular damage.

Patients should be reminded of the benign nature of the disease and informed that no further workup is necessary. Treatment is not required because Achenbach syndrome is self-limiting, but sargegrelate hydrochloride, acetysalicyclic acid, long acting diprimadol, heparin, isosorbidedinitrate, mucopolysaccharidepolysuplphate gel, and cold applications have been reported.[5] In our case, we provided DerMend bruise cream for the patient to use upon recurrence of symptoms.

CONCLUSION

Achenbach syndrome is a rare, benign condition marked by self-resolving hematoma on the palmar aspect of the digits. The underlying cause of this disease is unknown although genetic links to thrombotic pathways have been suggested. If Achenbach syndrome is diagnosed, physicians should avoid unnecessary testing and advise patients of its benign prognosis.

Conflict of Interest Disclosures: None

Funding: None

Corresponding Author:
Chelsea Taylor Schwartz, DO
Still OPTI/ Northeast Regional Medical Center
Email: chelseaschwartz@atsu.edu

References:

1. Godoy A, Tabares AH. Achenbach syndrome (paroxysmal finger hematoma). *Vasc Med*. 2019;24(4):361-366. doi:10.1177/1358863X19849627
2. Harnarayan P, Ramdass MJ, Islam S, Naraynsingh V. Achenbach's Syndrome Revisited: The Paroxysmal Finger Hematoma May Have a Genetic Link. *Vasc Health Risk Manag*. 2021;17:809-816. Published 2021 Dec 14. doi:10.2147/VHRM.S342847

3. Azarfar A, Beg S. Achenbach Syndrome: A Case Series. *Cureus*. 2022;14(3):e22824. Published 2022 Mar 3. doi:10.7759/cureus.22824
4. Gaurav V, Grover C. Achenbach Syndrome: A Rare Case of Paroxysmal Finger Hematoma. Indian Dermatol Online J. 2021;12(5):774-775. Published 2021 Aug 2. doi:10.4103/idoj.IDOJ_822_20
5. Ada F, Kasimzade F. Analysis of 24 patients with Achenbach's syndrome. World J Clin Cases. 2019;7(10):1103-1110. doi:10.12998/wjcc.v7.i10.1103
6. 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 Jun 27;40:1-2. doi: 10.1016/j.ejvssr.2018.05.008.
7. Harnarayan P, Ramdass MJ, Islam S, Naraynsingh V. Achenbach's Syndrome Revisited: The Paroxysmal Finger Hematoma May Have a Genetic Link. Vasc Health Risk Manag. 2021 Dec 14;17:809-816. doi: 10.2147/VHRM.S342847.
8. Ashrafzadeh S, Xue Y, Horn TD, Korman JB. The case of a painful blue thumb. JAAD Case Rep. 2020 Jul 31;6(10):1053-1055. doi: 10.1016/j.jdcr.2020.06.045.