

Acute Non-Ischemic Idiopathic Blue Finger: Achenbach's Syndrome. Case Report and Literature Review

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ABSTRACT

Achenbach's Syndrome (AS) is a pathology with low incidence and its etiology is unknown. It is characterized by the acute appearance of blue coloration in the finger without a triggering event and might be confused with an ischemic event. However, AS is a self-limited disease without sequelae. This case report is about a 75-year-old woman who presented sudden onset pain and purple coloration in the index finger of her right hand. Diagnostic tests were unhelpful and did not provide relevant information. She received symptomatic treatment, achieving complete resolution. The anamnesis and an exhaustive physical examination play a primary role in the suspicion; leaving diagnostic tests for the exclusion of other pathologies when they are required. We believe that knowing about this pathology allows a correct approach leading to an appropriate diagnosis.

Keywords: Achenbach's syndrome; digital bruising; blue finger; paroxysmal haemorrhage.

Level of Evidence: IV

Dedo azul agudo idiopático no isquémico: síndrome de Achenbach. Presentación de un caso y revisión bibliográfica

RESUMEN

El síndrome de Achenbach es un cuadro de baja incidencia y de etiología idiopática. Se caracteriza por la aparición aguda de coloración azul digital sin un episodio desencadenante, imitando a un cuadro isquémico, pero, al contrario, el síndrome de Achenbach se autolimita sin dejar secuelas. Se presenta una mujer de 75 años que consultó en la central de emergencias por dolor y cambio de coloración violácea del dedo índice de su mano derecha, de inicio súbito. Los estudios complementarios no aportaron información relevante. Se administró un tratamiento sintomático y se logró la resolución completa del cuadro. La anamnesis y el examen físico exhaustivo tienen un rol fundamental para sospechar este cuadro y los estudios complementarios se reservan para descartar otras enfermedades, si es necesario. Consideramos que conocer esta enfermedad y un abordaje correcto conducen a un diagnóstico apropiado.

Palabras clave: Síndrome de Achenbach; hematoma digital; dedo azul; hemorragia paroxística.

Nivel de Evidencia: IV

INTRODUCTION

Achenbach's syndrome, also called blue finger syndrome, paroxysmal finger hematoma, or acute idiopathic blue finger, is scarcely described in the literature.¹⁻⁶ Although a low incidence is reported—up to five cases per million inhabitants—its diagnosis could be underestimated.^{4,7-9} It has a higher prevalence in the female population between 40 and 60 years of age.^{2,7,10,11} Although its etiology is unknown, it has been linked to spontaneous subcutaneous hemorrhage and various hypotheses on the alteration of vascular flow and capillary fragility have been postulated to explain this phenomenon.^{2,4,5,7,9,12-20} As its presenting clinical characteristics suggest an ischemic episode,

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knowledge of this condition should be more widespread, taking into account that patients receive medical attention through different specialties, such as dermatology, vascular surgery, internal medicine, family medicine, plastic surgery, and orthopedics. It is important to know about this condition mainly to avoid expensive and invasive diagnostic studies and to be able to offer adequate treatment and follow-up.^{9,11,13,21}

The objective of this article is to present a case of Achenbach's syndrome and a literature review, proposing a simple and safe diagnostic route.

CLINICAL CASE

A 75-year-old retired woman with a dominant right hand. She reported a history of polymyalgia rheumatica, hypothyroidism, depression, dyslipidemia, and chronic gastritis. She was taking aspirin 100 mg/day. She attended the Orthopedic Emergency Department due to the sudden appearance of a purplish color on the index finger of her right hand. She reported that pain and edema had spontaneously appeared two hours earlier. After a few minutes, she noticed color changes on her finger. It turned purplish, accompanied by a sensation of heat and functional limitation. She did not mention a triggering cause and denied having experienced trauma or exposure to sudden changes in temperature. She had not suffered similar episodes before.

Upon physical examination, a purplish coloration of the volar region of the index finger was observed, without the involvement of the distal phalanx. It had increased in diameter compared to the contralateral finger and flexion was limited by edema (Figure 1).



Figure 1. Clinical image of the right hand. **A.** Palmar view. **B.** Dorsal view.

The following was found: capillary refill time <2 seconds, adequate local temperature, palpable peripheral radial and ulnar pulses, negative Allen test at the wrist and finger levels. Sensitivity was preserved, with two-point discrimination <6 mm. Pain and functionality were assessed using the visual analog scale (0 = no pain; 10 = worst pain) and the DASH score (0 = best result, 100 = worst result). The score for pain was 5/10 at rest and 7/10 while active, and the baseline DASH score was 63.75. Radiographs did not reveal bone involvement (Figure 2).



Figure 2. Radiograph of the right index finger. **A.** Anteroposterior. **B.** Lateral.

In turn, due to soft tissue involvement, an ultrasound and Doppler ultrasound were urgently requested, which showed subcutaneous edema, preserved arterial flow, and absence of thrombosis. Biochemical analyzes were performed that included hemogram, coagulogram (platelet count, prothrombin time, activated partial thromboplastin time), ionogram, erythrocyte sedimentation rate, C-reactive protein, and lipid profile. All of these values were within normal parameters. Taking into account both the clinical evaluation and the complementary tests, the patient was indicated to rest the hand and take oral analgesia as needed.

During the successive controls, a progressive decrease in clinical signs was observed, the condition healed after two weeks without leaving sequelae, and there were no recurrences until the final discharge four months after onset (Figure 3). At that time, the evaluation yielded a score of 0/10 for the visual analog scale both at rest and in activity and a DASH score of 16.5.



Figure 3. Clinical image of the right hand. **A.** Palmar view. **B.** Dorsal view.

DISCUSSION

The first report of this condition dates back to 1958, by Walter Achenbach.²² It is more prevalent in white ethnic groups, the female sex (90%), and during the middle age.^{3,4,7,10,23} Despite that the etiology is not clear, Robertson et al. associate this condition with a reduction in finger flow demonstrated by angiography; conversely, Takeuchi et al. do not report changes in finger flow.^{3,17} With the use of capillaroscopy, hemorrhages were observed, without morphological changes in the capillaries. On the other hand, the fragility of the small vessels was postulated, possibly related to age and the hormonal factor.^{8,16,18} In addition, Kämpfen et al. published that the use of ergotamine can cause a vasospastic reaction.¹² As numerous studies assure, this would not be an expression of a systemic disease, but it is associated with rheumatoid arthritis, autoimmune thyroiditis, gastrointestinal and gallbladder disease, acrocyanosis, and migraine.^{1,14,24}

Its clinical presentation consists of the sudden appearance of a blue-violaceous color change, frequently preceded by prodromal symptoms, such as itching, discomfort, paresthesia, or pain.^{2,4,25} After the color change, which may turn pale or reddish, the symptoms that accompany the condition are pain (60-100%), edema (60%), paresthesia (20-40%), functional limitation, and, to a lesser extent, a sensation of heat or coldness.^{7,10,11,14,17,23,24} The color change is explained, both by histology and by imaging studies, as a subcutaneous hemorrhage mechanism that does not go through the common stages of a hematoma.^{1,3,4,15,16,19} Its onset is considered to be spontaneous, although some reports associate it with mechanical phenomena, such as minor trauma, shearing effects during repetitive activities with the affected hand, and sun exposure.^{3,7,11,16,19} The most affected region is the hand, predominantly on the right side, and the index and middle fingers, but it can appear in other areas, such as the feet.^{2,7,10,11,15,24} The absence of involvement of the distal phalanx and its delimitation to the volar region is characteristic, although it is not a pathognomonic sign.^{2,4,21} According to some authors, there is a risk of recurrence (an average of up to 3 episodes) and, in other publications, the presence of a family member with a history of Achenbach's syndrome was associated as a predisposing factor.^{7,9,23} Treatment is purely symptomatic, with analgesics,

cryotherapy, immobilization, and rest.^{4,7} Its prognosis is benign, it does not leave sequelae, and it resolves spontaneously within a few days of the onset, although it can last up to six weeks, according to some authors.^{2,3,7,21}

The physical examination is essential to assess the different differential diagnoses. The affected areas should be evaluated taking into account the following: if the entire finger is involved, an ischemic episode or Raynaud's syndrome may be suspected; if acral areas are affected, acrocyanosis, acorhigosis, or pernio should be suspected; and if it appears in regions other than the acral ones, it can be considered psychogenic purpura (due to emotional stress) or dermatitis artefacta.^{7,21} The affected finger may be cold in the ischemic episode or Raynaud's syndrome, or hot in erythromegaly.²¹ Cutaneous stigmas, such as ulcers or gangrene, are observed in thromboangiitis obliterans, and nodules (thrombi) are present in digital vein thrombosis.^{7,21} The palpation of peripheral pulses and Allen test, performed bilaterally on both the wrist and the fingers, are useful when an ischemic episode of embolic, thrombotic, vasospastic or vasculitic origin is suspected.¹¹ The Adson test is usually positive in thoracic outlet syndrome.^{7,21} Finally, the presence of systemic signs and symptoms that may suggest collagen disease or bacterial endocarditis must not be disregarded.^{7,11,21}

As it has been proven, the complementary methods employed do not provide specific positive data for the diagnosis of Acherbach syndrome and their usefulness lies in ruling out differential diagnoses.^{2,4,7,24} They include measuring blood pressure with the ankle-brachial index, pulse oximeter saturation, conventional ultrasound, and venous and arterial Doppler ultrasound. Other studies include laboratory tests, such as hemogram, coagulogram, international normalized ratio, ionogram, kidney and liver function, erythrocyte sedimentation, C-reactive protein, lipid and immunological profile (antinuclear antibodies, rheumatoid factor, anti-DNA), viral serology, radiographs, MRI, echocardiogram, angiotomography or angiography, capillaroscopy, and even a biopsy.^{3,9,12,16-19,21} If necessary, noninvasive methods, such as comprehensive laboratory tests, radiography, and Doppler ultrasound, should first be used to rule out traumatic injuries, immune and vascular disorders. If the diagnosis cannot be established, it is possible to advance to more invasive studies such as angiography or angiography, which can be useful both for diagnosis and for eventual treatment.

We consider that the lack of knowledge about Acherbach syndrome and the belief that it represents an emergency condition reveal the importance of its awareness. For this reason, we believe that an exhaustive anamnesis and a correct physical examination would be sufficient for its diagnosis and, therefore, complementary tests should be reserved for cases of diagnostic doubt.

Conflict of interest: The authors declare they do not have any conflict of interest.

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