



Achenbach Syndrome: A Benign Painful Blue Finger with Tip Sparing

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Vascular surgeons are often consulted for patients with spontaneous painful discoloration of fingers and toes. In most cases, no diagnosis can be ascertained after extensive investigations and the condition resolves spontaneously. Awareness of Achenbach syndrome among physicians may help mitigate anxiety in patients because it is relatively benign and has a good prognosis. This report presents a case of Achenbach syndrome in an Asian woman along with a literature review.

Key Words: Achenbach syndrome, Vascular disease, Trauma

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INTRODUCTION

Achenbach syndrome, also known as “painful blue finger” or “paroxysmal finger hematoma,” is a rare clinical condition, which results in the sudden onset of bruising along with burning pain, mostly on the volar aspects of fingers. Knowledge regarding this syndrome can assist in optimal diagnosis and, thus, help alleviate anxiety because the syndrome is relatively benign and has a good prognosis. This report presents a case of Achenbach syndrome in an Asian woman along with a literature review.

CASE

A 56-year-old female visited our clinic presenting with sudden-onset edema and bruising in the right third finger (Fig. 1). Five years earlier, the patient had experienced a similar episode that subsided spontaneously within a week without any treatment. She was previously diagnosed with breast cancer, which treated with surgical resection and hormonal therapy. She did not have any specific medical history, including a history of vascular medicine or cyanosis of the hands and feet. Laboratory test results including

rheumatologic screening (such as rheumatoid factor, anti-nuclear antibody, ANCA, and ASO titer) were normal. Similarly, coagulation test results did not reveal any questionable findings. The patient’s condition gradually improved and her symptoms were completely resolved within two weeks without any specific treatment. Based on the clinical presentation and benign course, a diagnosis of Achenbach syndrome was made.

DISCUSSION

Since the German physician Walter Achenbach originally described Achenbach syndrome in 1958 [1], only around 100 cases of Achenbach syndrome have been published thus far, with a majority of them from the west. Since sporadic cases of symptoms in the toes have been reported, the term “paroxysmal acral hematoma” implies the fact that both fingers and toes may be affected. A French study indicated that the condition is actually more common, with the prevalence of 12.4% in women and 1.2% in men, and the age of onset is predominantly over 50 years in the general population. The third finger of the right hand is the most frequently involved area. Recurrence is common and the



Fig. 1. Characteristic bruise of Achenbach syndrome in the third finger of the right hand (A, dorsal surface; B, volar surface).

mean number of episodes is 3.04 [2]. In general, the distribution of discoloration is limited to the volar aspect of the digit, with sparing of the fingertip.

The etiology of Achenbach syndrome remains unclear, but intermittent spontaneous hematoma formation in the volar surface of the hands is a characteristic symptom. One of the distinctive features is that the hematological variables, including platelets and clotting factors, are within normal values. Symptoms are secondary to infiltration and compression effect of extravasated blood.

It is important to differentiate between Achenbach syndrome and other similar conditions as these symptoms and signs may suggest a more serious vascular disease, which may require invasive investigations.

Acute limb ischemia should be differed as it demonstrates discoloration, pain, edema, and paresthesia. The differentiating factors are female predominance and normal peripheral temperature in the case of Achenbach syndrome. Furthermore, time to resolution of the symptoms is limited to a few weeks. This condition is restricted to individuals aged

<60 years of age; however, acute limb ischemia can affect any age group. Raynaud's syndrome has similar recurrent episodes, but the pale finger manifestation is aggravated by cold exposure. Thromboangiitis obliterans (Buerger disease) resembles acute limb ischemia with a similar presentation, although it is mainly associated with tobacco use and results in eventual ulceration and gangrene. Associated conditions include acrocyanosis, gastrointestinal diseases, migraine, and gall bladder disease; however, the pathophysiologic relationship is unclear. Another similar condition is "painful bruising syndrome," which is a rare but distinctive clinical entity, thought to be due to allergic sensitivity to red cells in the tissues. It is characterized by a distinctive localized purpuric reaction occurring primarily on the legs, face, and trunk, with recurring painful ecchymoses variably accompanied by syncope, nausea, vomiting, and gastrointestinal and intracranial bleeding. Dermatitis artefacta is a rare self-induced psychocutaneous disorder, in which mechanical and chemical devices are most commonly used to produce injuries. In most cases, the patient denies his/her role in its causation. Hence, it is different from Achenbach syndrome.

In addition to providing the clinical course description, some authors have performed specific examinations. Layton and Cotterill [3] reported that H&E staining was unhelpful in confirming a definitive diagnosis by a biopsy of an affected area of skin and that stains for amyloid were negative. It has been hypothesized that in some patients, increased capillary resistance and vascular fragility may trigger Achenbach syndrome even after minimal trauma. A capillaroscopic study of the affected finger showed multiple severe hemorrhages without further alterations of capillary morphology or blood flow [4]. Robertson et al. [5] demonstrated decreased blood flow to the metacarpophalangeal joint of the affected digit, suggesting vasospastic disorder. Therefore, current diagnosis is usually made based on the clinical assessment and exclusion of similar hematologic disorders. Awareness of specific disease characteristics, such as spontaneously resolving recurrent bruise in the finger, are important, and one can suspect this syndrome by obtaining a thorough history from the patient.

Complete resolution usually occurs within a few days, but symptoms may last for a few months. Recurrent episodes occur for a variable period of time (months or years) without any apparent lasting sequelae. Because of its benign nature, no specific prevention and treatment has been proposed [6].

This rare case of Achenbach syndrome in an Asian woman suggests that as the clinical course of Achenbach syndrome is relatively benign with a good prognosis, optimal diagnosis and reassurance is crucial. There is no need for unnecessary invasive investigations.

CONFLICTS OF INTEREST

The author has nothing to disclose.

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