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CASE REPORT

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Achenbach Syndrome – Case Report and Discussion on the Interdisciplinary Approach of a Patient

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ABSTRACT

We present a case of Achenbach syndrome, which is an alarming disease for both patients and physicians, although it is a rare, benign and self-limiting disorder. It is also reported as "paroxysmal finger haematoma", but the majority of cases are misdiagnosed, and unnecessarily investigated and treated.

Keywords: Achenbach syndrome, bluish discoloration fingers, acute ischemia.

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INTRODUCTION

Achenbach syndrome is an alarming disease for both patients and physicians, although it is a rare, benign and self-limiting disorder. It is also reported as "paroxysmal finger haematoma", but the majority of cases are misdiagnosed and unnecessarily investigated and treated. The recognition of the disease, mainly on clinical features, is the purpose of the study. ❑

CASE REPORT

A 68-year-old Caucasian female patient was seen in Emergency for sudden onset of bluish discoloration of the volar area of her left hand and digits I, II, III. At the time of examination, she reported tingling sensation but no pain or pruritus. The patient denied previous exposure to cold, emotional stress, or any other possible trigger factors (Figures 1, 2). She was very anxious when admitted to the Cardiology Unit for a suspicion of thromboembolic event or acute ischemia. On clinical examination, all extremities were warm and peripheral pulses palpable. Extensive laboratory investigations, including coagulation tests and inflammatory profile, were within normal limits. She remained hospitalized for three days under strict surveillance. She was stable and neither cardiac nor rheumatologic conditions were identified. Due to the suspicion of acute ischemia of the limb, Doppler sonography was performed but it revealed no pathological findings. In the absence of any topical or systemic treatment, the patient has completely recovered within 72 hours. Skin lesions have completely disappeared in less than three days, without any scar or skin discoloration. A retrospective diagnosis of Achenbach syndrome was done and the patient was reassured about the benign nature of the disease. ❑

FIGURE 1. Bluish discoloration of the volar area of left hand and digits I, II, III

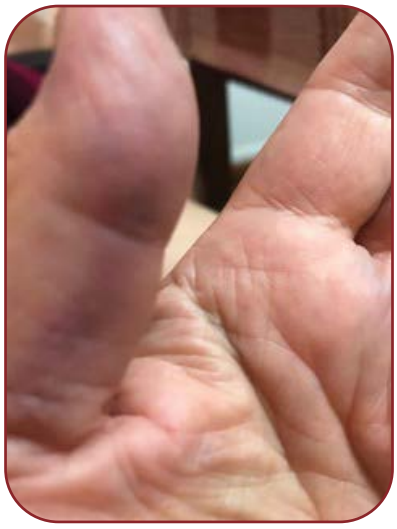

FIGURE 2. Bluish discoloration of the volar area of left hand and digits I, II, III

DISCUSSION

Since its first description in 1958, by Walter Achenbach (1), few cases have been reported, the disease being ignored or misdiagnosed. A retrospective systematic literature review of the syndrome revealed almost annually reports, with only one case in most reports, quantifying less than 100 cases in English database (2).

The clinical image catches the eye and becomes unforgettable once seen. The first description, made by Achenbach, is still used without revisions nowadays, being clear and useful for diagnosis. The clue of the diagnosis is based on unique or recurrent episodes of sudden onset of pain and blue discoloration on the palmar areas of one or more digits (1, 2). Skin manifestations are observed on the volar surface of the hand fingers, mostly the index and middle finger, and on the palm. The disease affects mostly women of middle age and, characteristically, it resolves spontaneously within hours or days. No scars or

other remaining manifestations have been observed after each episode.

There are authors who have named the disease "paroxysmal hand hematoma" or "finger apoplexy", but it was recommended to maintain its initial name for a better recognition all over the world and avoidance of misinterpretation of the disease (3).

The pathogeny of the disease is still unknown, although many theories have been launched, including thromboembolic or atheroembolic incidents, capillary microhaemorrhages or vasculitis (4). No causative relation with local trauma, exposure to heat or cold, physical effort have been proved and patients do not recall any identifiable trigger factor.

All cases are initially seen in the Emergency or Cardiology Units for a presumptive diagnosis of digital venous thrombosis or acute ischemia and extensive investigations are performed, with all proving to be within normal limits. Then, patients are referred to Rheumatology, Hematology or Vascular departments for further investigations for new suspicions such as Raynaud syndrome, acrocyanosis, and collagen vascular diseases (5).

The term syndrome was primarily use to describe the symptomatology, while the etiology was unclear. However, we do not know much more than Achenbach in 1958. The best concept is that of a vasomotor disease. ❑

CONCLUSIONS

The disease worries both patients and physicians, but it should be kept in mind that it is a benign condition with spontaneous resolution and no requirements for expensive and invasive investigations and treatments. ❑

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