Aïnhum (Dactylolysis Spontanea), A Rare Mutilant Disease : A Case Report

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Abstract

Aïnhum, also known as dactylolysis spontanea, is a rare dermatological disease that most frequently affects men of African origin, characterized by a fibrous constrictive band that develops preferentially at the base of the 5th toe. The natural evolution of aïnhum can be described in four different stages, ranging from a simple fibrous band to self-amputation of the toe. The classic distinction is between "true" aïnhum of unknown cause, which would only affect people of African descent, and pseudo-aïnhum, which follows various causes. We present a case of aïnhum, at stage 2 of the pathology, treated with a Z-plasty on the 5th toe. The diagnosis is clinical, and the prognosis after early surgical treatment is favourable.

Keywords: Aïnhum; Dactylolysis Spontanea; Spontaneous Amputation

Introduction

Aïnhum, or dactylolysis spontanea, is a rare condition that produces constricting rings by a fibrous band around a toe, or more rarely a finger, leading after several years to spontaneous amputation. The provenance of the root «aïnhum» is uncertain but seems to mean fissure in the language of the Nagos tribe of Brazil. It's also possible that it's related to «ayun», the Lagos tribe’s word for saw, as the two tribes share a similar ancestor [1]. Although extremely rare, the condition is a traumatic and painful experience that can be prevented by early diagnosis.

Case Report

A 76-year-old female from Togo was referred to the Dermatology Department with a painful deformity of the 5th toe of the right foot. She had presented two years earlier with a fissure at the same level. On clinical examination, there was a hyperkeratotic ring constriction at the base of the toe and swelling of the distal part. Her collateral and hereditary history consists only of hypertension treated with potassium-sparing diuretic. An ultrasonography showed no collection or formation suggestive of a tumor mass (Figure 1). The patient was referred to the Department of Orthopedic Surgery. Radiographs and a magnetic resonance imaging of the toe showed tissue stricture with osteoporosis of the bone pieces (Figure 2 and 3). After comparison of the various examinations, the diagnosis of aïnhum was retained. A Z-plasty was performed on the lateral and medial borders to free the fibrous bands (Figure 4). The postoperative course was simple, the patient’s skin progressed favorably, and her pain disappeared (Figure 5). Seven months after the operation, there was still some stricture on the medial side of the toe. A topical retinoid treatment was initiated but no regression was obtained after three month. A second Z-plasty procedure was carried out only on the medial side. The patient was followed up for a possible early diagnosis of contralateral involvement.
Figure 1: Ultrasonography of the 5th toe of the right foot: the formation at the level of the last phalanx corresponds to a fleshy-looking tissue. No cyst or mass was identified.

Figure 2: Radiographic image of the right foot (A) and comparatively of the left foot (B). There is tissue stricture at the base of the 5th toe of the right foot and bone resorption at this level. Dysmorphia is also seen contralaterally.

Figure 3: T1-weighted magnetic resonance imaging of the right foot in coronal (A), sagittal (B), and transverse (C) sections. There is no demonstrated mass syndrome, no signal abnormality of the bony elements or tendon injury. Sequences were performed after gadolinium injection but did not provide any new information.

Figure 4: Medial view of the 5th toe of the right foot. The Z-plasty is performed, allowing the release of the band. A second plasty is performed laterally.

Figure 5: Preoperative (A) postoperative (B) and 1-month (C) appearance of the 5th toe of the right foot, showing a reduction in annular constriction as well as a reduction in distal edema.

Discussion

The term “aïnhum” was introduced in 1867 by Da Silva Lima who described the existence of constrictive bands in the Brazilian population of African descent [2]. Aïnhum, also known as dactylolysis spontanea, presents with a progressive development of a circumferential fibrous band most frequently at the base of the 5th toe, leading to self-amputation after 4-5 years [3]. The disease can affect any phalanx (finger or toe) and bilateral forms in the feet are frequently encountered (75% of cases). It occurs mainly in people aged between 20 and 50 years with black skin, from Africa, but also from Western India and South America. Men are more prone than women with a sex ratio of 2:1. The prevalences in the literature are 2.2%, 0.2% and 0.0015% respectively in Nigeria, Congo and Panama [4].
In 1952, a classification was proposed by Wells, et al. according to the following designations: aïnhum, congenital bands, aïnhu-moid bands associated with other diseases, and bands secondary to trauma [5]. Neumann, et al. proposed the term «pseudo-aïnhum» in 1953 to classify the latter three categories and clearly distinguish them from aïnhum. The authors also emphasized that the amniotic band syndrome does not fall under the designation of pseudo-aïnhum but is a differential diagnosis in newborns [6].

By definition, «true» aïnhum is a constrictive band without an identifiable cause. In contrast, pseudo-aïnhum results from an identifiable or associated pathologic process and is independent of skin color. Pseudo-aïnhum is not limited to the extremities and can be seen on the entire body. This differentiation has not always been respected, resulting in misclassification of cases reported in the literature [1].

Pseudo-aïnhum therefore includes a number of forms. These include hereditary disorders of keratinization with palmoplantar keratoderma, including Vohwinkel syndrome (hereditary mutilating keratoderma). Infectious causes such as mycotic infections, leprosy, schistosomiasis, syphilis or yaws have been described. Pseudo-aïnhum also occurs in systemic diseases with vascular and articular tropism, such as systemic or localized scleroderma, rheumatoid arthritis, systemic lupus, as well as in diseases with advanced peripheral neuropathy and/or vascularization disorders of the extremities. Diabetes and alcoholism are the main contributors. Some traumatic cases have been observed in children by wrapping tissue fibers, hair or elastic around a toe (tourniquet syndrome) [3,7].

The etiology of true aïnhum remains unclear to date. In the past, factors related to the rural lifestyle in Africa have been incriminated. Currently, their responsibilities have been ruled out as the condition is observed regardless of social status. Genetic factors have also been proposed, based on the association of pseudo-aïnhum with hereditary syndromes such as Vohwinkel syndrome [8]. According to Browne, et al. aïnhum is considered to be a primitive disorder of the skin based on a congenital tendency to excessive fibroblastic response to various stimuli, especially to trauma and chronic infections [9].

Radiological imaging is useful to identify the extension of the disease. Indeed, Cole, et al. describe four stages according to clinical and radiological features. In stage I, a fold develops and progresses into a groove or fissure surrounding the toe. In stage II, the toe becomes globular distally, associating arterial narrowing and bone resorption. In stage III, very painful, the bone separates at the joint with hypermobility of the toe. Stage IV consists of non-bloody self-amputation [10]. Although biopsies are not required to make the diagnosis, there is localized dermal fibrosis replacing muscle tissue, sweat glands, and blood vessels. Nerves are usually intact [11]. Electron microscopic study shows the same features found in keloid tissue [12].

There are no guidelines for the management of the condition. The treatment for stage I and II is the same as for scars and constrictions on the extremities. The use of corticosteroids, retinoids or salicylates in the form of injections or topicals is mentioned in the literature. Complete disappearance of the constrictive bands has been observed in several publications after 1 to 2 months of oral vitamin A treatment. Surgically, resection of the fibrous band, Z-plasty, cross-finger flap (temporary pedicle flap taken from the lateral-dorsal aspect of the adjacent finger), or skin grafting may prevent progression of the disease. Once stages III and IV are reached, the possible outcomes are surgical amputation or acceptance of spontaneous amputation of the affected extremity [1,9,13].

Conclusion
Aïnhum is a pathological process whose etiology remains unknown. It evolves in four stages from circumferential cracking of the toe to spontaneous amputation. The observations presented in this article remind us that it is important to know the signs of aïnhum, as well as to keep in mind all the syndromes that can be associated with it. A simple surgical treatment allows to quickly relieve the pain while preserving the affected toe and avoiding the psychological and mutilating trauma.

Bibliography


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