

Pseudoainhum Associated With Lepromatous Leprosy: A Very Rare Association in Modern World

Ahangama Arachchige Nilanga Nishad^{1,*}, Mahathevan Pathman², Seyed Ahamed Uwyse³, Ashan Mendis³, Prabath Kularathne Abeysundara⁴, Arjuna P. de Silva⁵

¹ Professorial Medical Unit, Colombo North Teaching Hospital, Colombo 11010, Sri Lanka; ² Department of Dermatology, National Hospital of Sri Lanka, Colombo 01000, Sri Lanka; ³ Department of Dermatology, Colombo North Teaching Hospital, Colombo 11010, Sri Lanka; ⁴ Department of Clinical Medicine, General Hospital, Ampara 32350, Sri Lanka; ⁵ Department of Clinical Medicine, Faculty of Medicine, University of Kelaniya, Kelaniya 11300, Sri Lanka.

Abstract

Introduction: Pseudoainhum (dactylolysis spontanea) is characterized by the development of a fibrous band around the digit that gradually leads to autoamputation. Digital pain associated with Pseudoainhum may not be evident in patients with neuropathic conditions.

Here, we present a rare case of pseudoainhum patient, which describes a very rare association of pseudoainhum with leprosy.

Case presentation: A 48 year old male with lepromatous leprosy, with resorption of digits, charcot joints and tropical ulcers was seen in the clinic. The fourth digit of the left hand had a narrowing due to a fibrous band at the 2nd inter phalangeal joint with shiny tethering distal phalanx. He could not recall the duration of the ainhum. He was not suffering from any pain.

Discussion: The current report describes a very rare association of pseudoainhum with leprosy. The case involved a 48-year-old man in the dermatology ward with lepromatous leprosy presenting with a pseudoainhum.

Conclusion: This case highlights the possibility of delayed presentation of patients with pseudoainhum to physicians when the patients have underlying neuropathic conditions that prevent feeling finger pain and cause abnormal appearance of the digits, especially in leprosy.

Keywords: pseudoainhum leprosy, lepromatous leprosy, case report

Introduction

Pseudoainhum or ainhum (dactylolysis spontanea) is an acquired or congenital disorder, characterized by the development of a fibrous band around the digit that gradually leads to autoamputation. Ainhum on the other hand is the idiopathic condition which was initially described as a form of leprosy but was later considered a different condition.¹ Pseudoainhum is known to be associated with diabetes, syphilis, porphyria, scleroderma, syringomyelia,

and atypical keratoderma. If identified in the early stages, treatments to relieve the constriction can be offered, thus preventing autoamputation of the digit. One of the clinical presentations of pseudoainhum is digital pain.² However, associated conditions such as lepromatous leprosy or diabetic neuropathy may allow the pseudoainhum to progress to the end stage of autoamputation without pain. Additionally, patients with leprosy may not recognize the abnormal digit appearance caused by pseudoainhum because of confusion with lepromatous digit changes that have already taken place. As a result, the patient will not present to the health care provider for relief of the constriction in the initial stages of the disease.

Here, we present a rare case of pseudoainhum patient, which describes a very rare association of pseudoainhum with leprosy.

Case report

A 48-year-old man presented with numbness in his hands and feet of several years' duration. On examination, he had peripheral polyneuropathy in his upper and lower limbs. He also had resorption of the digits, and trophic ulcers were seen. The fourth digit of the left hand exhibited focal narrowing due to a fibrous band at the second

* Corresponding author: Dr. Ahangama Arachchige Nilanga Nishad, Professorial Medical Unit, Colombo North Teaching Hospital, Colombo, Ragama 11010, Sri Lanka. E-mail: aanilanga@gmail.com.

Conflicts of interest: The authors declare that they have no conflicts of interest

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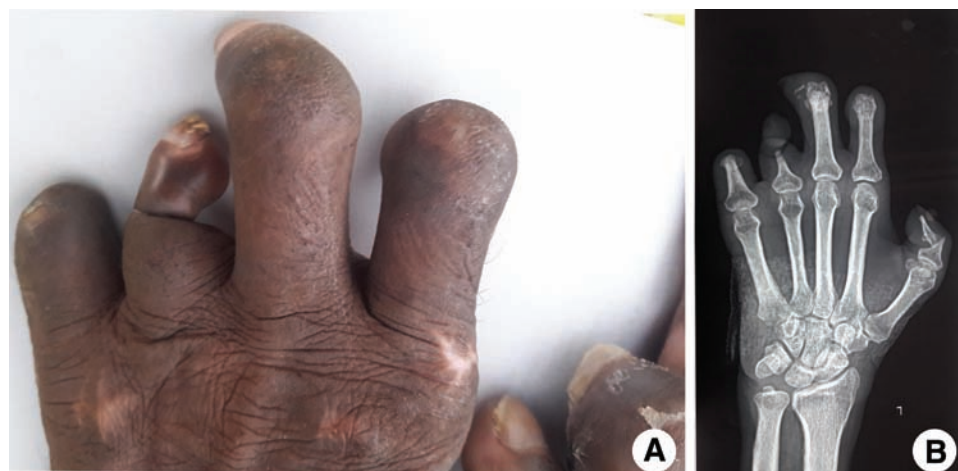


Figure 1. Clinical features of a patient with pseudoainhum and lepromatous leprosy. (A) Ainhum involving the fourth digit of the left hand in a patient with lepromatous leprosy. (B) Left hand X-ray image showing focal narrowing due to a fibrous band at the second interphalangeal joint with a shiny tethering distal phalanx.

interphalangeal joint with a shiny tethering distal phalanx (Fig. 1). He had several hypopigmented anesthetic patches in upper back of the chest and on the upper and lower limbs. He had no thickened nerves. He had a leonine-like face with bilateral ear lobe infiltration. A slit-skin smear from both ear lobes showed acid-fast bacilli. A skin biopsy from upper back showed granulomatous dermatitis with perineural elongated granulomas.

A diagnosis of pseudoainhum and lepromatous leprosy was made based on the clinical and histopathological results, and he began 12 months of multidrug therapy for multibacillary leprosy and was referred to a plastic surgeon for management of the constricting digital band. However, the patient was lost for follow-up after surgical referral.

Patient consent declaration

The patient gave the written informed consent about her case publication, including the images and other disease details.

Discussion

Pseudoainhum or ainhum (dactylyolysis spontanea) is caused by a fibrotic band that encircles a digit and constricts it, gradually resulting in autoamputation. The condition is most commonly seen among people in their second to fifth decades of life and is more prevalent among men.³ Ainhum is idiopathic, whereas pseudoainhum is always secondary.⁴ The incidence of pseudoainhum mentioned few decades ago in tropical and subtropical climates ranges from 0.015% to 2.0% of the population. The fibrotic band classically begins within a flexural groove. The lesion is circumferential and often slowly progresses with resultant changes distal to the constriction. It usually affects the fifth digit of the toe. Pseudoainhum is associated with many conditions, such as diabetes, syphilis, porphyria, scleroderma, syringomyelia, atypical keratoderma, and leprosy.

The clinical criteria for a diagnosis of ainhum/pseudoainhum include 3 findings: (1) soft tissue constriction; (2) distal bulbous enlargement; and (3) distal thinning or lysis of phalangeal bones. Additionally, it is divided into 4 clinical stages: (1) a clavus develops and progresses to an annular fissure around the finger; (2) the finger becomes globular distal to the groove because of lymphedema,

and this change is associated with bone resorption and arterial narrowing; (3) the bone painfully separates at the joint with hypermobility of the finger; and (4) severely painful bloodless autoamputation of the toe occurs.

Our patient had all of the clinical diagnostic features of pseudoainhum, including soft tissue constriction, distal bulbous enlargement, and distal thinning or lysis of the phalangeal bones.⁵ He was in the late stage of pseudoainhum with hypermobility of the fingers, but neuropathy of leprosy had rendered his hands painless. The patient also refused surgical interventions.⁶

In summary, this report describes a very rare association of pseudoainhum with leprosy because the patient did not have the common conditions that are usually associated with pseudoainhum. It also highlights the possibility of delayed presentation of patients with pseudoainhum to physicians if they have coincidental or etiologic neuropathic conditions that prevent them from feeling finger pain. Some patients may not experience pain in the initial stages but instead have only an “abnormal look” of the fingers; in such cases, confusing the abnormal appearance of the digits with leprosy may also delay the initial consultation. Even though we suggest the association of lepromatous leprosy for pseudoainhum, the rarity may affect the study on the generalizability of casual-relationship with it, which may be a major limitation of this case report.

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