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Unilateral Pseudo-Ainhum in Liver Cirrhosis

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Abstract

BACKGROUND: Pseudo-ainhum is defined as any case of auto-amputation not associated with the classic spontaneous ainhum seen in Africans with unknown etiology.

CASE PRESENTATION: A severely ill 58-year-old male patient presented with a painless constricting circular band on his left second toe. His medical history was remarkable for severe alcoholic liver cirrhosis with ascites formation leading to dyspnea. He had a hypoalbuminemia and a pronounced peripheral sensory neuropathy.

CONCLUSION: Here we present the second case of pseudo-ainhum associated with liver cirrhosis.

Introduction

Liver cirrhosis is considered as an end-stage of different types of liver injury. It is characterised by a chronic inflammatory and fibrotic process [1]. Cirrhosis has been associated to several skin diseases such as soft tissue infections [2], yellow urticaria [3], spider angiomas, paper money skin and xerosis [4], and Muehrcke lines of the nails [5].

In 2001, Wollina et al. described 64-year-old Caucasian woman with breast cancer, systemic scleroderma, and primary biliary cirrhosis due to Reynolds' syndrome, who presented with bilateral pseudo-ainhum [6]. Here, we report a second case of pseudo-ainhum in a patient with liver cirrhosis.

Case Presentation

A severely ill 58-year-old male patient presented with a painless constricting circular band on his left second toe. His medical history was remarkable for severe alcoholic liver cirrhosis with ascites formation leading to dyspnea. He had a hypoalbuminemia and a pronounced peripheral sensory neuropathy. Other comorbidities were hypertension and hyperuricemia.

On examination we observed a constricting band of the second left toe (Figure 1). He had a generalized xerosis cutis with features of paper money skin and purpura, but no jaundice. He had palmar erythema and onychomycosis of toe nails.

We made the clinical diagnosis of pseudo-ainhum stage I. The primary treatment consisted of the management of the underlying liver disease.

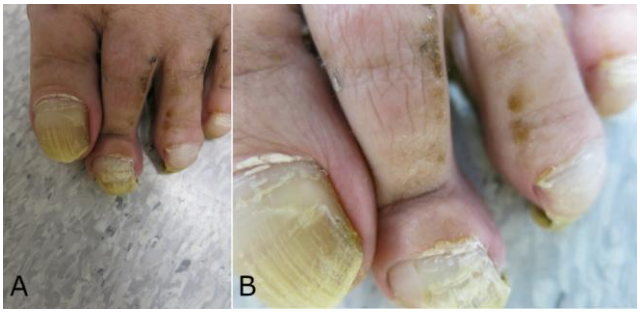

Figure 1: A) Constricting band of the 2nd toe on the left foot, onychomycosis; B) Detail demonstration the constricting circular fibrotic band, onychomycosis and hyperkeratosis of the cuticle

Discussion

Ainhum (dactylosis spontanea) is a rare mutilating disorder of fingers and toes, most frequently seen in Africans. Ainhum develops in four stages. In the beginning, a clavus progresses to an annular fissure or band around the digit. While the soft tissue constriction gets more pronounced, the digit becomes globular distal to the groove. This is associated with bone resorption and arterial narrowing. Stage 3 describes a very painful bone separation at the joint with hypermobility of the distal part of the digit. In stage 4 a bloodless auto-amputation of the toe happens, which is associated with severe pain [7].

The major differential diagnosis of ainhum is pseudo-ainhum. Pseudo-ainhum is defined as any case of auto-amputation not associated with the classic spontaneous ainhum seen in Africans with unknown etiology. Grading is similar to ainhum [8]. Pseudo-ainhum has been described secondary to congenital amniotic bands known as Streeter's syndrome [9] or keratoderma hereditarium mutilans (Vohwinkel) [10]. Pseudo-ainhum can also occur as a very rare complication of infectious diseases (lues, leprosy or yaws), ichthyosis, scleroderma, or ischemia [11], [12], [13]. This is the second report about pseudo-ainhum and liver cirrhosis [6]. In liver cirrhosis, liver parenchyma is replaced by excess extra-cellular matrix leading to tissue fibrosis. The pro-fibrinogenic cytokines platelet-derived growth factor and transforming growth factor-β1 stimulate the production of collagen, noncollagenous glycoproteins, proteoglycans, and glycosaminoglycans while matrix-metalloproteinases are downregulated [1]. Tissue fibrosis is also involved in pseudo-ainhum, but the target cells are skin fibroblasts instead of hepatic stellate cells and hepatic myofibroblasts.

Treatment guidelines do not exist for pseudo-ainhum. Dependent on the underlying pathology and stage, systemic retinoids or surgery have been reported [14], [15], [16]. In the present case, retinoids were contraindicated. Surgery was not recommended due to the hepatic impairment of blood coagulation and the general medical situation of the patient.

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