Unilateral Pseudo-Ainhum in Liver Cirrhosis

Uwe Wollina¹, Michael Tirant², Aleksandra Vojvodic³, Veronica di Nardo², Torello Lotti², 4

¹Department of Dermatology and Allergology, Städtisches Klinikum Dresden, Academic Teaching Hospital, Dresden, Germany; ²Department of Dermatology, University of Rome "G. Marconi", Rome, Italy; ³Military Medical Academy of Belgrade, Belgrade, Serbia; 4Hanoi Medical University, Hanoi, Vietnam

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.

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.
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 claw 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 profibrinogenic 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.

References

1. Romanelli RG, Stasi C. Recent advancements in diagnosis and therapy of liver cirrhosis. Curr Drug Targets. 2016; 17(15):1804-1817. https://doi.org/10.2174/1389450117666160613101413 PMid:27296314
2. Sanglodkar U, Jain M, Jothimoni D, Parida S, G B, Venkataraman J. Cellulitis in liver cirrhosis - a series of 25 cases from southern India. Clin Exp Hepatol. 2018; 4(3):201-204. https://doi.org/10.5114/ceh.2018.78125 PMid:30324146 PMCID:PMC6185931
3. Chiba T, Hayashi F, Shimura M, Kyomatsu M, Tatamatsu S, Nakao M, Funue M. Yellow urticaria in a patient with hepatic cirrhosis. G Ital Dermatol Venereol. 2014; 149(3):381-2.
4. Dogra S, Jindal R. Cutaneous manifestations of common liver diseases. J Clin Exp Hepatol. 2011; 1(3):177-84. https://doi.org/10.1016/S0973-6883(11)60235-1
5. Muehrcke RC. The finger-nails in chronic hypoalbuminaemia; a new physical sign. Br Med J. 1956; 1(4979):1327-8. https://doi.org/10.1136/bmj.1.4979.1327 PMid:13316143 PMCID:PMC1990060
6. Wollina U, Graefe T, Oelzner P, Hein G, Schreiber G. Pseudoainhum of all fingers associated with Reynolds’ syndrome and breast cancer: report of a case and review of the literature. J Am Acad Dermatol. 2001; 44(2 Suppl):381-4. https://doi.org/10.1067/mjd.2001.104971 PMid:11174422
7. Daccarett M, Espinosa G, Rahimi F, Eckerman CM, Wayne-Brunot S, Couture M, Rosenblum J. Ainhum (dactylosis spontanea): a radiological survey of 6000 patients. J Foot Ankle Surg. 2002; 41(6):372-8. https://doi.org/10.1016/S1067-4501(02)80083-7
8. Rashid RM, Cowan E, Abbasai SA, Brieva J, Alami M. Destructive deformation of the digits with auto-amputation: a review of pseudo-ainhum. J Eur Acad Dermatol Venereol. 2007; 21(6):732-7. https://doi.org/10.1111/j.1468-3083.2007.02224.x PMid:17567298
9. Piccolo V, Cornell P, Piccolo S, Zalaudek I, Argenziano G, Russo T. Pseudo ainhum and facial malformation secondary to Streeter's dysplasia. J Eur Acad Dermatol Venereol. 2019. https://doi.org/10.1111/jdv.15757 PMid:31222806
10. Bassetto F, Tiengo C, Serrazza R, Belloni-Fortina A, Alabacib M. Vohwinkel syndrome: treatment of pseudo-ainhum. Int J Dermatol. 2010; 49(1):79-82. https://doi.org/10.1111/j.1365-4632.2009.03467.x PMid:20465619
11. Behera B, Ghochait D, Thappa DM. Pseudoainhum and autoamputation associated with lamellar ichthyosis. Indian J Dermatol Venereol Leprol. 2017; 83(6):728-729. https://doi.org/10.4103/ijdvl.IJDVL_56_17 PMid:28936990
12. Silva M, Ribeiro R, Oliveira J. Pseudoainhum in systemic sclerosis. J Rheumatol. 2016; 43(11):2076. https://doi.org/10.3899/jrheum.160239 PMid:27803439
13. Dasan BV, McBrearty A, Lau L, Lee B. Pseudoainhum of the toe with underlying chronic lower-limb ischemia. Int J Low Extrem Wounds. 2011; 10(2):96-7. https://doi.org/10.1177/1534736111409369 PMid:21893445
14. Richey PM, Stone MS. Resolution of pseudoainhum with acitretin therapy in a patient with palmoplantar keratoderma and congenital alopecia. JAAD Case Rep. 2019; 5(3):219-221. https://doi.org/10.1016/j.jrcom.2018.12.004 PMid:30809564 PMCID:PMC6374960
15. Piso T, Bhattacharyya S, Oberlin C. Surgical correction of pseudo-ainhum in Vohwinkel syndrome. J Hand Surg Br. 1995; 20(3):338-41. https://doi.org/10.1016/0266-7817(95)00030-7
16. Damin M, Watson S. Loricrin palmoplantar keratoderma: full-thickness skin grafting for pseudoainhum. Clin Exp Dermatol. 2019; 44(4):444-446. https://doi.org/10.1111/ced.13770 PMid:30264492