Case Report

Dactylolysis Spontanea or Ainhum Involving the Big Toe

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ABSTRACT. We describe the case of an Italian Caucasian man with ainhum involving both big toes. Ainhum or dactylolysis spontanea is characterized by the development of a constricting band around a toe, which progresses to spontaneous autoamputation. It usually affects the fifth toe bilaterally, but in rare cases other toes may be involved. The disease occurs in Black people living in tropical regions but occasionally has been reported in persons having fair skin. (J Rheumatol 2005;32:2437–9)

Key Indexing Terms:
AINHUM DACTYLOLYSIS SPONTANEA

Ainhum, also known as dactylolysis spontanea, is a distinct clinical and radiological disorder of dark-skinned peoples characterized by the development of a progressive deepening and constricting band that encircles the toe and usually results in spontaneous bloodless amputation[1-5]. It most typically affects both little toes, but in rare cases other toes may be involved[6]. Pain is a common feature and shows variable intensity. Both sexes are affected, but it is slightly more common in men. Occasionally ainhum has been reported in persons having fair skin[6]. We describe an Italian Caucasian man suffering from dactylolysis spontanea of both toes.

CASE REPORT

A 58-year-old man was referred to us in January 2003 for evaluation of a 2-year history of painful fissure in his right big toe (Figure 1). Seven years before, the same symptoms and signs had affected the left big toe. An infection developed and the toe was amputated. On examination his right big toe had a groove beginning from the plantar digital fold and encircling nearly all its circumference. The toe was held dorsiflexed and rotated laterally. His history was otherwise unremarkable.

Routine laboratory evaluation was normal. Radiographs showed a constricting radiolucent band around the base of his right big toe together with a marked narrowing of the proximal phalanx, more prominent on the medial aspect (Figure 2). Magnetic resonance imaging revealed the constricting band of soft issue that was more prominent on the medial side (Figure 3). There was no involvement of flexor and extensor tendon insertions. Arteriography of both feet was normal (Figure 4).

DISCUSSION

Our patient showed 2 atypical aspects of ainhum: being Caucasian and the fact that the big toe was involved. Nevertheless, the clinical and radiological findings were typical of the disease. Other conditions that may produce constricting bands and mimic ainhum were excluded.

The term "ainhum" is derived from a word meaning "saw" in the African Yoruba dialect. The disease occurs in Black people living in Africa, Asia, South and Central America, and the United States[1-5]. The greatest incidence values have been found in Nigeria, with 2.48 per 1000 men and 1.06 per 1000 women[3,4]. Ainhum has been considered a very rare disease in African Americans. Only 30 cases of ainhum had been reported in the North American literature until 1992[7]. Recently, Daccarett, *et al*[1] reviewed 4126 consecutive radiographic studies of feet performed in the African American population of Chicago. After an initial screening based on the Cole criteria for ainhum[2], 581 patients were selected and reexamined with enhanced visualization techniques for soft tissue. After review of the complementary examinations, 102 patients (1.7%) were diagnosed as having ainhum. The authors concluded that dactylolysis spontanea is underdiagnosed and overlooked in the African American population because of its low prevalence and variable clinical presentations that mimic more common etiologies including trauma, arthritis, and tenosynovitis.

Four clinical and radiological stages have been identified[2,3]. In stage I ("grooved digit"), a deep, soft tissue groove with hyperkeratotic debris is visible. It begins along the medial aspect of the distal portion of the proximal phalanx of the little toe and afterwards extends laterally. The constriction is usually narrow but may be sufficiently wide to give an "hourglass" aspect to the involved toe. The bone is not involved in this stage. In stage II ("bulbous digit"), a painful bulbous enlargement appears due to lymphedema of

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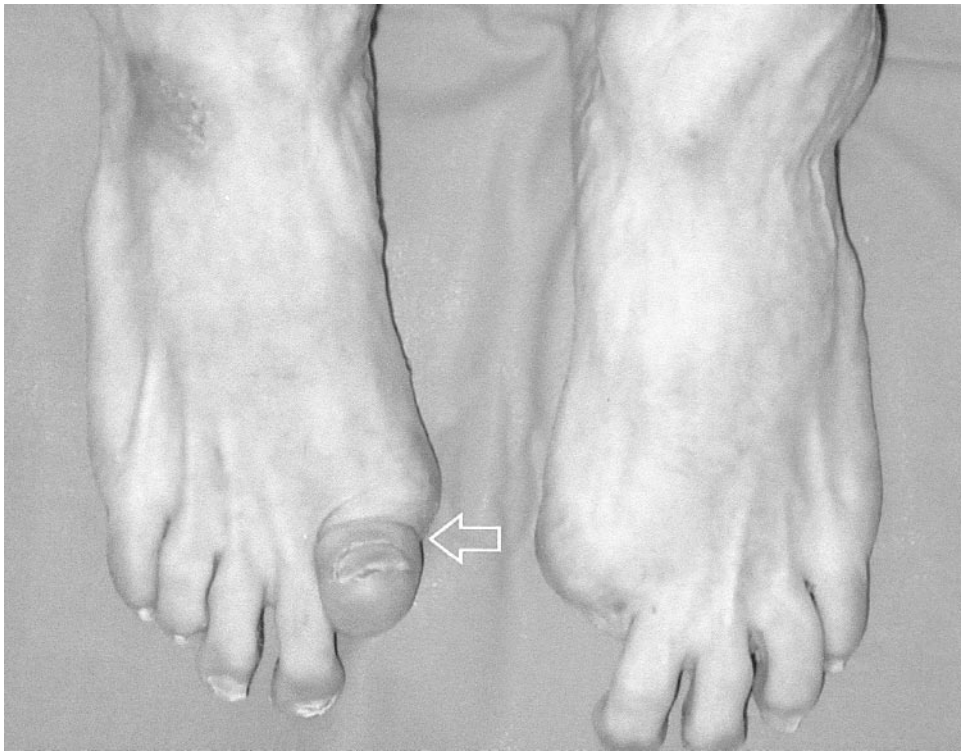


Figure 1. A groove beginning from the plantar digital fold and encircling the right big toe is visible (arrow). The left big toe was amputated previously.

Figure 2. Anteroposterior view of the right toe showing severe narrowing of the proximal phalanx, especially on the medial aspect, underlying the soft tissue groove (arrow).

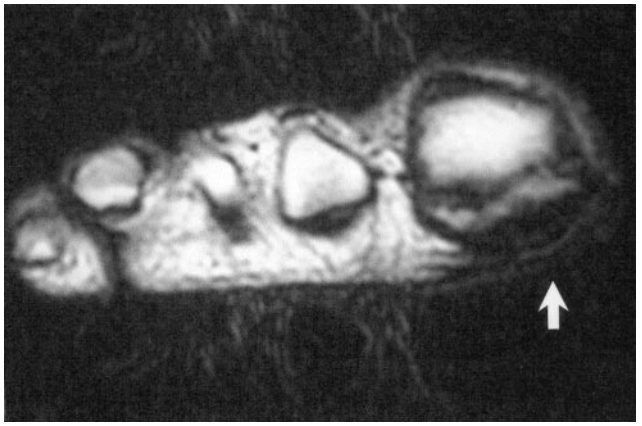
the toe distal to the constriction. Often there is an external rotation of the digit so that the nail faces laterally. Stage III ("dangling digit") is characterized by bone resorption that begins medially at the distal portion of the proximal phalanx. The phalanx gradually narrows with the progression of the resorption and eventually fractures with minimal trauma. Stage IV ("lost digit") is the spontaneous amputation of the dangling digit, which typically occurs with little pain and bleeding. The remaining stump has a unique triangular "iceberg" shape. The process typically develops over 4–5 years. However, superimposed aggressive infections may hasten the pathological process, and autoamputation may occur in a few months.

Patients in stages I and II ainhum may benefit from intralesional steroid injections, trauma prevention, surgical shoes, and Z-plasty with resection of the constricting band[8–10]. Once the bony changes appear, the prognosis for saving the toe becomes poor and surgical amputation of the toe may be necessary to relieve pain.

Histological examination in ainhum shows marked hyperkeratosis and acanthosis of the epidermis overlying chronic inflammation and fibrosis in the dermis and hypodermis[11].

The cause of ainhum is unknown. Many etiological factors have been suggested including hereditary predisposition, trauma, leprosy, yaws, syphilis, filariasis, schistosomiasis, fungi, and disorders of peripheral vessels and nerves[1].

Ainhum must be differentiated from other ainhum-like annular constricting bands (pseudoainhum), which may be



A

B

Figure 3. Axial T1 weighted fast-spin echo (A) and T2 weighted fast-spin echo (B) images show the constricting band of soft tissue (arrows).

Figure 4. Arteriography of the right foot showing normal plantar arch and its branches.

congenital or associated with other acquired disease entities including disorders of keratinization, syringomyelia, diabetes mellitus, infection (syphilis, leprosy), trauma (burn, frostbite), and connective tissue diseases (localized scleroderma, systemic sclerosis, discoid lupus erythematosus)[12-15]. In none of these conditions is there the predilection for the medial aspect of the distal part of the proximal phalanx of the little toe typical of aihnum.

Our case reminds rheumatologists of the existence of this poorly recognized disease.

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