A Rare Etiology of Nontraumatic Recurrent Leg Ulcers: Prolidase Deficiency

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Abstract

Prolidase deficiency is an autosomal recessive disorder. It has many clinical manifestations such as mental retardation, chronic skin ulcers, recurring infections, splenomegaly and characteristic facial appearance. We present a case of prolidase deficiency, a 42 years old woman who have many bilateral chronic leg ulcers, which firstly occurred when she was 6 years old. She also have other clinical components of prolidase deficiency. Laboratory analysis revealed an elevated erythrocyte sedimentation rate, hypergammaglobulinemia, and iron deficiency anemia. A large amount of hydroxyproline was detected after hydrolysis of a 24-h urine specimen (99 mg/day).

Keywords: Leg ulcers, Prolidase deficiency,

Introduction

Prolidase deficiency is an autosomal recessive and multisystemic disorder [1]. It should be think when if there are chronic, non-healing skin ulcers.

Case Study

A 42 years old woman who have many bilateral chronic leg ulcers, which firstly occurred when she was 6 years old. The patient had experienced numerous deep polygonal ulcers which are with elevated bordes. There was edema and induration on both of legs and the ulcers located especially on the back side of the legs and dorsal face of feet. Also some atrophic lesions are detected on this area. There was similar disease on her family history. In addition to this skin lesions, the patient has the other components of prolidase deficiency such as, splenomegaly and chronic otitis media, mental retardation, deafness. Physical examination showed a characteristic facial appearance which include hypertelorism, depressed nasal root and micrognathia. Laboratory analysis revealed hypergammaglobulinemia, anemia and increasing hydroxyproline levels in urine (99mg/day). She treated with systemic and topical antibiotics, vitamin C, then it is started to hydrocolloid dressing treatment.

Discussion

Prolidase deficiency is an autosomal recessive disorder [1,2]. It is firstly described by Goodman in 1968 [1,3]. Prolidase is an enzyme that divides iminodipeptides such as proline and hydroxyproline [1,2]. Prolidase deficiency patients has iminopeptiduria due to absence of prolidase enzyme activity, so it results some clinical manifestations such as mental retardation, chronic skin ulcers, recurring infections, splenomegaly and characteristic facial appearance, e.g saddle nose, frontal bossing, hypertelorism, micrognathia, thick lips [1,2].

Figure 1: Characteristic facial appearance; hypertelorism, depressed nasal root, micrognathia.

Figure 2: Chronic leg ulcers; chronic, nonhealing, recurring leg ulcers on the back side.
Protuberant abdomen, keratosis pilaris, photosensitivity, dry skin deafness, osteoporosis, joint laxity, scaly erythematous maculopapular rash, hypertrichosis, lymphedema also can be observed [1,2]. Coexistence of prolidase deficiency and lupus erythematosus has been described [1,4-6]. There is no treatment to completely heal but there is some helpful options such as vitamin c and manganese, systemic or topical antibiotics, pulse corticosteroid treatment [1,7-9].

**Conclusion**

Prolidase deficiency is a rare disease but it should be think as a reason of the chronic and recurring skin ulcers.

**References**