A Case of Acrokeratoelastoidosis

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A 28-year-old woman had a 13-month history of skin-colored, well-circumscribed, hyperkeratotic plaques on both heels. Histological examination showed hyperkeratosis and acanthosis in the epidermis. Special stain for elastic tissue revealed marked fragmentation, diminution and thickening of elastic fibers in the dermis. We report a rare case with the lesions of acrokeratoelastoidosis on both heels. (Ann Dermatol 13(2) 126-128, 2001).

Key Words: Acrokeratoelastoidosis

Acrokeratoelastoidosis (AKE) is a rare skin disorder originally described by Costa in 1953 which is inherited by autosomal dominant, but may be sporadic. To our knowledge, only six cases of AKE have been reported in Korea\(^1\) (Table 1), but no case with the lesions of AKE on the heel has previously been reported in Korea. We report the first case with the lesions of AKE on both heels in Korea.

CASE REPORT

A 28-year-old woman presented with several plaques on both heels. The lesion began 13 months ago and was asymptomatic. Family history was negative for similar skin lesions. Clinical examination showed well-circumscribed, skin-colored hyperkeratotic and smooth surfaced confluent plaques on the medial and lateral regions of both heels (Fig. 1).

Histological examination revealed hyperkeratosis and acanthosis in the epidermis (Fig. 2). No specific changes were detected in the dermis in hematoxylin and eosin stain.

Elastin stain (Verhoeff-van Gieson) revealed marked fragmentation and diminution of elastic fibers in the reticular dermis (Fig. 3(A)). Some of the fragmented elastic fibers appeared thickened in the deeper portions of the dermis(Fig. 3(B)). Based upon these findings, a diagnosis of AKE was established. Because the skin lesion was asymptomatic and caused no great cosmetic problem, we did not give her any treatment.

DISCUSSION

Acrokeratoelastoidosis (AKE) was described by Costa in 1953 as the familial or sporadic appearance of keratotic papules and plaques on the hands and feet\(^3\). It is clinically characterized by firm, shiny papules on the lateral margins of the palms and soles\(^3\). The lesions most commonly appear during childhood or adolescence and usually remain throughout life. They are most often asymptomatic.

Histologically, the essential feature consists of hyperkeratosis, acanthosis, diminution and fragmentation of elastic fibers\(^5\). Some of the fragmented elastic fibers appear thickened and tortuous\(^5\).

AKE belongs to the complex group of disease defined as marginal papular acrokeratodermas\(^7\) (table 2). Marginal papular acrokeratodermas share keratotic papules, usually crateriform, along the border of the hands and feet as a common clinical finding. All these diseases therefore need to be differentiated from AKE. This is usually done on the basis of differences in the

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Fig. 1. Skin lesions of the medial region of the right heel (A) and the lateral region of the left heel (B).

Fig. 2. Hyperkeratosis and acanthosis in the epidermis. (H&E, ×10).

According to clinical and histological features. Especially, the differential diagnosis between AKE and focal acral hyperkeratosis (FAH) is difficult because both conditions have very similar clinical and histological manifestations, but fragmentation and diminution of the elastic tissue are observed in AKE, whereas the elastic tissue is intact in FAH. Generally, treatment is not necessary, but there was a report that etretinate was used successfully.

REFERENCES


Fig. 3. Elastin stain shows marked fragmentation and diminution of elastic fibers (A), and thickening of some fragmented elastic fibers (B). (Verhoeff-Van Gieson, ×400).
Table 1. Summary of reports of AKE in Korea

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Age/Sex</th>
<th>Onset</th>
<th>Mode of transmission</th>
<th>Site</th>
<th>Treatment</th>
<th>Response</th>
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<tbody>
<tr>
<td>1</td>
<td>75/M</td>
<td>Adult</td>
<td>Sporadic</td>
<td>Hands</td>
<td>TA injection</td>
<td>Good</td>
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<tr>
<td>2</td>
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<tr>
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<td>Sporadic</td>
<td>Hands</td>
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<tr>
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<td>52/F</td>
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<td>Hands</td>
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</tr>
<tr>
<td>5</td>
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<td>Infancy</td>
<td>AD</td>
<td>Hands</td>
<td>Topical tretinoin &amp; oral etretinate</td>
<td>Poor</td>
</tr>
<tr>
<td>6</td>
<td>20/M</td>
<td>Infancy</td>
<td>AD</td>
<td>Hands</td>
<td>Topical tretinoin &amp; oral etretinate</td>
<td>Poor</td>
</tr>
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<td>Our patient</td>
<td>28/F</td>
<td>Adult</td>
<td>Sporadic</td>
<td>Feet</td>
<td>No</td>
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AD, Autosomal dominant. TA, Triamcinolone acetonide

Table 2. Classification of marginal papular acrokeratodermas

<table>
<thead>
<tr>
<th>Hereditary with elastorrhexis</th>
<th>acrokeratoelastoidosis</th>
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<td>Hereditary without elastorrhexis</td>
<td>focal acral hyperkeratosis</td>
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<td>hereditary papulonodular keratoderma</td>
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<tr>
<td>Hereditary papulotranslucent keratoderma</td>
<td>acrokeratoderma hereditarium punctatum</td>
</tr>
<tr>
<td>Hereditary mosaic acral keratosis</td>
<td>mosaic acral keratosis</td>
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</table>

Acquired degenerative collagenous plaques of hands

digital papular calcinosis