

# Acrokeratoelastoidosis of Costa: A Case Report

## AKROKERATOELASTOİDOZ: OLGU SUNUMU

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### Abstract

Acrokeratoelastoidosis of costa is a rare papular palmoplantar keratosis in which small, round, firm, yellowish papules are seen on the margins of the hands and feet. The onset of the skin lesions is often before the age of 20. Both autosomal dominant transmission and sporadic occurrence have been reported. Characteristic histologic findings are marked hyperkeratosis, epidermal hypertrophy and the hallmark is the fragmentation and the decrease in number of the elastic fibers in the dermis. We report a case of acrokeratoelastoidosis in a 18-year-old Turkish girl and a review of the literature.

**Key Words:** Acrokeratoelastoidosis, akrokeratoderma, elastorrhexis

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### Özet

Akrokeratoelastoidoz el ve ayak kenarları boyunca küçük, sert, yuvarlak, sarımsı papüllerin bulunduğu nadir bir papüler palmoplantar keratozdur. Deri lezyonlarının başlangıcı sıklıkla 20 yaşından önce gelir. Hem otozomal dominant geçiş hem de sporadik oluşum bildirilmiştir. Karakteristik histolojik bulgular belirgin hiperkeratoz, epidermal hipertrofi ile dermiste elastik liflerin sayısında azalma ve fragmantasyondur. Bu makalede akrokeratoelastoidoz tanısı alan 18 yaşında bir kadın olgu sunulmakta ve konu ile ilgili literatür gözden geçirilmektedir.

**Anahtar Kelimeler:** Akrokeratoelastoidoz, akrokeratoderma, elastoreksis

**A**crokeratoelastoidosis (AKE) is a rare skin disorder which was first described by Oswaldo Costa, a Brazilian dermatologist, in 1953.<sup>1</sup> Clinically, small, firm, yellowish white, translucent, sometimes umblicated and/or hyperkeratotic papules are seen on the hands and feet. The most characteristic locations of these papules are the thenar and hypothenar areas and the boundary between the dorsal and palmar or plantar skin. The knuckles and nail folds may also be involved. The papules may coalesce to form plaques on the dorsal surfaces and the knees.<sup>2,3</sup> Although the lesions are generally asymptomatic, hyperhidrosis may be present.<sup>1,4,5</sup> Herein we report a case of sporadic AKE in an 18-year old patient. As far as

we know, this is the fourth case reported in our country.

### Case Report

An 18-year-old Turkish girl was seen in December of 2004 for a contact eczema on her hands and asymptomatic, small, firm papules which had been present since the age of 7 were noticed. Her contact eczema resolved after treatment with topical corticosteroid in one week and she came again to the clinic for evaluation of multiple papules on her hands and feet. The lesions first developed at about the age of 7 years and they increased progressively in number over the years. They were neither painful nor pruritic, and there was no evidence for hyperhidrosis. There was no history of excessive sun exposure or trauma. She was treated with a variety of preparations containing salicylic acid several times without any improvement.

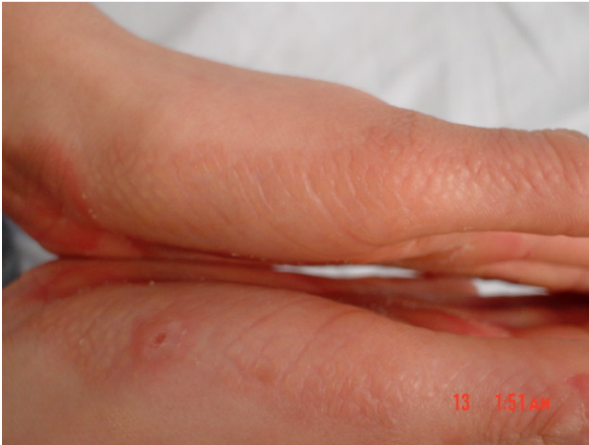
On physical examination, there were small (2-3 mm in diameter), firm, flesh-colored, oval and

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**Figure 1.** Multiple round and oval, yellowish-white, firm papules along the boundary between the dorsal and palmar surfaces of both hands.



**Figure 2.** Numerous small flesh-colored keratotic papules coalescing into plaques on the dorsal surfaces of interphalangeal joints.

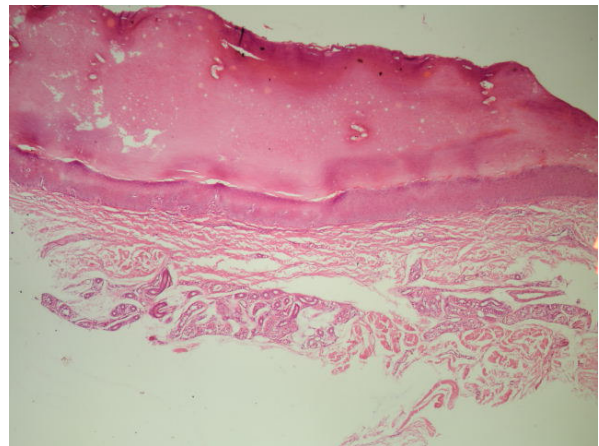
round multiple papules on both palmar, thenar and hypothenar regions and over the dorsal aspects of metacarpophalangeal and interphalangeal joints (Figure 1, 2). Similar papules were also present on the medial aspects and plantar surfaces of both feet (Figure 3). There were no other similar lesions on the other parts of the patient's body.

A biopsy specimen was obtained from one of the papules on the thenar margin of the left hand for histologic examination. On hematoxylin-eosin staining, epidermal hyperplasia, hyperkeratosis and a prominent granular zone were shown (Figure 4) and elastic tissue stain (Verhoff stain) revealed

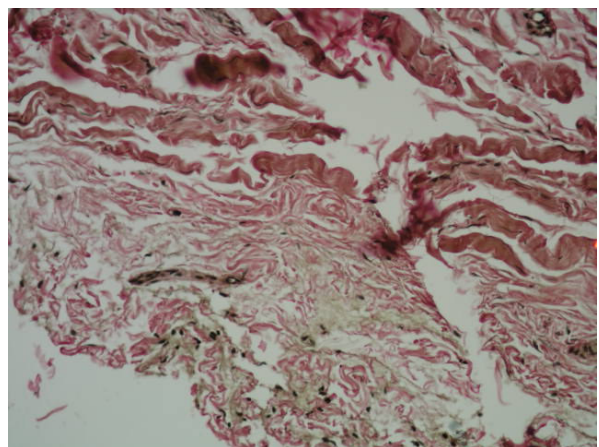
qualitative diminution and fragmentation of elastic fibers in the underlying dermis (Figure 5).



**Figure 3.** Similar lesions along the medial aspect of right foot.



**Figure 4.** Epidermal hyperplasia, hyperkeratosis and a prominent granular zone (Hematoxylin-eosin stain; magnification X 60).



**Figure 5.** The elastic fibers in the dermis are decreased in number and are fragmented (Verhoff elastic tissue stain; magnification X 200).

**Table 1.** The differential diagnosis of acrokeratoelastoidosis.<sup>14</sup>

Disorder	Age of Onset	Race and Sex	Etiology	Clinical Features	Histologic Features
Acrokeratoelastoidosis (AKE)	childhood adolescence	none	Idiopathic genetic (OD)	Small, round-oval to rhomboid shaped, yellowish papules on borders of palmar / plantar surfaces	Hyperkeratosis, epidermal hypertrophy, decreased and fragmented elastic fibers in the dermis (elastorrhexis)
Focal acral hyperkeratosis (FAH)	childhood adolescence	females black	Idiopathic genetic	Identical to AKE	Same as above except elastorrhexis in the dermis, elastic tissue is intact
Degenerative collagenous plaques of the hands (DCP)	adulthood	males caucasoid	UV pressure	Keratotic papules on radial borders of hands	Marked degeneration of collagen and elastic fibers, senil elastosis
Keratoelastoidosis marginalis of the hands (KEM)	adulthood	males australians	UV pressure	Keratotic papules on the radial margin of the index finger and the opposing margin of the thumb	Identical to DCP
Mosaic acral keratosis (MAK)	youngsters	females Negroid	idiopathic genetic	Keratotic papules over finger joints, hyperkeratosis of palms and knees	Hyperkeratosis, acanthosis, no hypergranulosis, normal elastic fibers

### Discussion

Acrokeratoelastoidosis of Costa is characterized by small, yellowish white, firm papules with dermal histologic marker elastorrhexis, located on the margins of the hands and feet.<sup>3</sup> The age of onset is usually childhood or early adolescence, and the disease usually progresses slowly over years.<sup>4,6</sup> A rapid progression has been reported during pregnancy.<sup>4</sup> The lesions of our patient were first appeared at the age of 7 and the distribution of lesions was typical, they were located along the borders of the hands and feet predominantly.

Histologically, the hallmarks of AKE are marked hyperkeratosis, epidermal hypertrophy and decreased numbers of elastic fibers, with coarse fragmentation in the dermis.<sup>2,3,7</sup> The histopathological examination of our patient's biopsy material revealed hyperkeratosis and characteristic fragmentation of elastic fibers. Electron microscopy studies have also shown this reduction in numbers and morphologic abnormalities of the elastic fibers. Dense granules in or near the plasma membrane of fibroblasts have been detected on electron microscopy by Masse et al.<sup>5</sup> Based on these findings, it has been suggested that there may

be a defect in the secretion or excretion of elastic fibers from fibroblasts.<sup>5,8</sup> But the definitive cause remains unknown. The disease is both reported to have an autosomal dominant inheritance and to be sporadic.<sup>4,7-12</sup> In 1962, Costa<sup>9</sup> himself reported in his monograph the occurrence of similar lesions in ten members of a family, and in 1974 Jung et al.<sup>10</sup> studied twenty-one cases in a large family through three generations and suggested an autosomal dominant inheritance pattern for the disease. A possible linkage to chromosome 2 has been suggested.<sup>13</sup> Our patient is accepted to be sporadic because any other member of her family had similar lesions.

AKE is included in the marginal papular acrokeratodermas which is a complex group of disorders that share keratotic papules along the borders of the hands and feet as a common clinical feature. Rongioletti et al.<sup>14</sup> propose the simplified concept of marginal papular acrokeratodermas that may be divided into the hereditary type with elastorrhexis (AKE), or without it (Focal acral hyperkeratosis), and the acquired type (Degenerative collagenous plaques). The keys for differential diagnosis of these conditions are summarised in Table 1.

Focal acral hyperkeratosis (FAH) was first described by Dowd et al.<sup>15</sup> in 1983. He studied 15 patients clinically identical to AKE but on histologic examination only focal hyperkeratosis and acanthosis were present, elastic stains were normal and the collagen appeared normal. FAH is a more common disorder of focal hyperkeratosis and occurs more commonly in blacks.<sup>2,3,14</sup>

Degenerative collagenous plaques (DCP) of the hands was first described by Burks et al.<sup>16</sup> in 1959 in a group of older patients. This skin disorder is seen in older Caucasian patients and is usually associated with prolonged sun exposure. The margins of the thumb, the index finger, ulnar sides of the hands, the middle and right fingers are often involved with asymptomatic, keratotic, yellowish papules. Histopathologically, active degeneration of collagen and elastic fibers is seen. Dense collagen bundles are mixed haphazardly with elastic tissue in most of the reticular dermis.<sup>2,7,14</sup>

Keratoelastoidosis of marginalis (KEM) of the hands was first described by Koscard<sup>17</sup> in 15 patients in 1964. It is also seen in elderly patients with a history of manual work. Clinically it is similar to AKE but the distribution of lesions is slightly different. In KEM the radial margin of the index finger and the opposing margin of the thumb are often involved, AKE has more widespread distribution, including the dorsal aspects of the hands, fingers, the lateral aspects of the feet and over the lower legs. Histologically, thickened collagen fibers and increased elastotic material in the upper dermis are characteristic for KEM.

DCP and KEM are accepted to be variants of the same entity. These disorders are easily distinguished from AKE with the age of onset, history of chronic sun exposure or manual work, clinical distribution of the lesions and histologic changes.

Mosaic acral keratosis (MAK) differs from AKE by the lack of elastic fiber changes in the dermis and clinically striking mosaic or jigsawpuzzle pattern of keratotic papules.<sup>18</sup> It is accepted to be a variant of FAH.<sup>14</sup>

Besides these marginal papular acrokeratodermas, the differential diagnosis of AKE should

also include palmar xanthomas, porokeratosis and verrucae planae.<sup>2,3</sup>

There is no effective treatment for AKE. Multiple therapies, such as liquid nitrogen, salicylic acid, tretinoin and prednisone have been tried with minimal success.<sup>6</sup> Since the lesions of AKE are asymptomatic and there have been no reports of malignant degeneration, no treatment is offered.

There are three cases of AKE reported in our country.<sup>19,20</sup> Our patient is the fourth case according to the Turkish literature. Doğan et al.<sup>19</sup> used acitretin treatment in their two cases but did not succeed. Erbil et al.<sup>20</sup> reported one case treated with adaphalene gel without any response. Our patient was treated with topical combination of salicylic acid and corticosteroid but no improvement was observed.

AKE of Costa is a very rare skin disorder and our patient is a case of sporadic AKE displaying the characteristic clinical and histopathological features of the disease. Since the disease is rare and the definitive cause is unknown, there are limited therapies with minimal success. However, as the number of reported cases is increased and more detailed etiopathogenetic investigations are performed in the future, new and more effective therapeutic options will be found and used.

## REFERENCES

1. Costa OG. Acrokeratoelastoidosis. *Dermatologica* 1953; 107:164-8.
2. Nelson- Adesokan P, Mallory SB, Lombardi C, Lund R . Acrokeratoelastoidosis of Costa. *Int J Dermatol* 1995;34: 431-3.
3. Hu W, Cook TF, Vicki GJ, Glaser DA. Acrokeratoelastoidosis. *Pediatric Dermatology* 2002;19:320-2.
4. Highet AS, Rook A, Anderson JR. Acrokeratoelastoidosis. *Br J Dermatol* 1982;106:337-44.
5. Masse R, Quillard A, Hery B, Toudic Le Her G. Acrokerato-elastoidose de Costa. *Ann Derm Venereol* 1977;104:441-5.
6. Odom RB, James WD, Berger TG. *Andrews' diseases of the skin*. 9<sup>th</sup> ed. Philadelphia: WB Saunders Company; 2000. p.249.
7. Korc A, Hansen RC, Lynch PJ. Acrokeratoelastoidosis of Costa in North America. *J Am Acad Dermatol* 1985;12: 832-6.
8. Johansson EA, Kariniemi AL, Niemi KM. Palmoplantar keratoderma of punctate type: Acrokeratoelastoidosis Costa. *Acta Derm Venereol* 1980;60:149-53.

9. Costa OG. Acroceratoses (Ceratodermias palmo-plantares). Impr Univ. Minas Gerais, Edit., Belo Horizonte, Brazil, 1962.
10. Jung EG, Beil FU, Anton-Lamprecht I, et al. Acrokeratoelastoidosis. *Hautarzt* 1974;25:127-33.
11. Matthews CNA, Harman RRM. Case report. Acrokeratoelastoidosis in a Somerset mother and her two sons. *Br J Dermatol* 1977;97 (suppl 15):42-3.
12. Saruk M, Taylor MR, Rudolph RI, et al. Acrokeratoelastoidosis. *Cutis* 1983;32:248-51.
13. Greiner J, Krüger J, Palden L, et al. A linkage study of acrokeratoelastoidosis, possible mapping to chromosome 2. *Hum Genet* 1983;63:222-7.
14. Rongioletti F, Betti R, Crosti C, Rebora A. Marginal papular acrokeratoderma: A unified nasography for focal acral hyperkeratosis, acrokeratoelastoidosis and related disorders. *Dermatology* 1994;188:28-31.
15. Dowd PM, Harman RRM, Black MM. Focal acral hyperkeratosis. *Br J Dermatol* 1983;109:97-103.
16. Burks JW, Wise LJ Jr, Clark WH. Degenerative collagenous plaques of the hands. *Arch Dermatol* 1960;82: 362-6.
17. Koscard E. Keratoelastoidosis marginalis of the hands. *Dermatologica* 1965;131:169-75.
18. Jacyk K, Smith A. Mosaic acral keratosis. *Clin Exp Dermatol* 1990;15:361-2.
19. Doğan G, Oram Y, Hazneci E, Müezzinoğlu B, Aydın A. Two cases of acrokeratoelastoidosis: Failure of acitretin treatment. *Turk J Dermatopathol* 1996;3-4:173-6.
20. Erbil H, Akar A, Taştan HB, Özcan A, Gür AR. Acrokeratoelastoidosis. *Turk J Dermatopathol* 2000;9:142-4.