A Case with KID Syndrome: Keratitis-Ichthyosis-Deafness

KID Sendromlu Bir Olgu: Keratit-İktiyoz-Sağırlık

Ülker GÜL, MD,[a]
Arzu KILIÇ, MD,[a]
Müzeyyen GÖNÜL, MD,[a]
Seray KÜLCÜ ÇAKMAK, MD,[a]
Tuba ÜNAL, MD[b]

[a]2nd Dermatology Clinic,
[b]1st Pathology Clinic,
Ankara Numune Education and Research Hospital, Ankara

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Yazışma Adresi/*Correspondence:*
Arzu KILIÇ, MD
Ankara Numune Education and Research Hospital,
2nd Dermatology Clinic, Ankara, TÜRKİYE/TURKEY
kilicarzu2004@yahoo.com

ÖZET Keratit- iktiyoz-sağırlık (KID) sendromu sağırlık, iktiyoziform eritroderma ve keratitle karakterize ektodermin nadir görülen konjenital bir hastalığıdır. Bildirilen olguların çoğunluğu sporadiktir. Bu sendromun klinik özellikleri iktiyoz ve sağırlığı içermektedir. Keratit kısmen geç görülen bir bulgudur ve bazen görülmeyebilir. Bu yazıda iktiyozu, sağırlığı, bilateral ekstropiyon ve blefariti olan bir olgu sunuyoruz.

Anahtar Kelimeler: Kelimeler: keratit; sağırlık; iktiyoz; blefarit; ekstropiyon

ABSTRACT Keratitis-ichthyosis-deafness (KID) syndrome is a rare congenital disorder of the ectoderm characterized by deafness, ichtyosiform erythroderma and often keratitis. The majority of the cases reported previously were sporadic. The clinical features of this syndrome include ichthyosis, deafness. Keratitis is rather a late finding and sometimes may not be seen. In this report, we present a patient who had ichtyosis, deafness, bilateral ectropion and blepharitis.

Key Words: Keratitis; deafness; ichthyosis; blepharitis; ectropion

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Keratitis-ichthyosis-deafness (KID) syndrome is a rare disorder characterized by keratitis, ichthyosis and neurosensorial deafness. Keratitis may be a late finding and in some cases, it may not be seen. In this report, a patient who did not have keratitis, but had blepharitis, ectropion and photosensitivity is reported.

CASE REPORT

A 34-year old man presented with the skin lesions that had been present since birth. In detailed examination, we have learnt that during the first year of his life, he developed figurate red-brown hyperkeratotic plaques. There were periodic exacerbations and remissions, but complete clearing of the skin was never noted. Scalp hair, eyebrows, eye lashes and body hair had always been sparse. Deafness and photophobia were added to these symptoms in the following years. He had had recurrent bacterial skin and eye infections. Intellectual development was normal. There was no consanguinity in the family and the other members of the family were normal.

On dermatological examination, the skin was dry, there were figurate hyperkeratotic patches over the body (Figure 1, 2). The hair was short, dry, and sparse. The eyebrows and eyelashes were scarce (Figure 3).

No pathology was detected in the routine laboratory examinations . Chest X ray graphies , abdominal and pelvic ultrasonographies were normal. The histopathological examination of the biopsy specimen of the skin revealed ichthyosis (Figure 4).

The audiogram of the patient revealed bilateral deafness of sensorineurial type. The patient was consulted with the opthalmologist for the photophobia and was diagnosed as blepharitis and ectropion.

DISCUSSION

Characteristic features of KID syndrome include an ichtyosiform dermatosis, neurosensory deafness, and keratitis.[1,2] Additional findings include sparse hair, and sparse or absent eyebrow and lashes, leukonychia, or thick dystrophic nails, oral mucosa and tongue involvement, dental anormalities.[3-5] The mode of inheritance is unclear but it is thought to be sporadic.[1,3] Recently, it has been related with the mutations in the connexin-26 gene.[4] Also familial cases have been reported.[1]

Generalized ichtyosiform erythroderma is usually present at birth or appears in infancy. In most cases sharply demarcated hyperkeratotic pla-

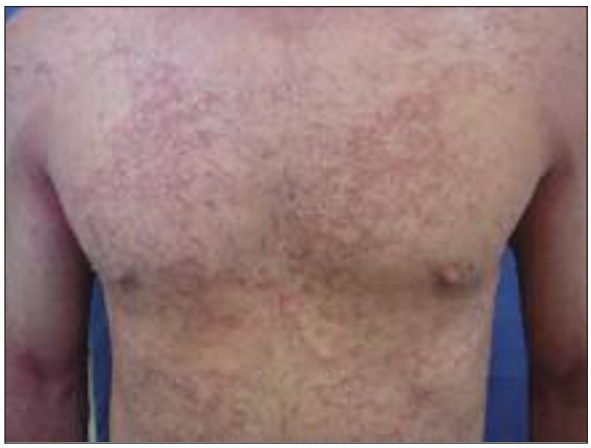

FIGURE 1: Figurate erythematous hyperkeratotic patches over the chest.

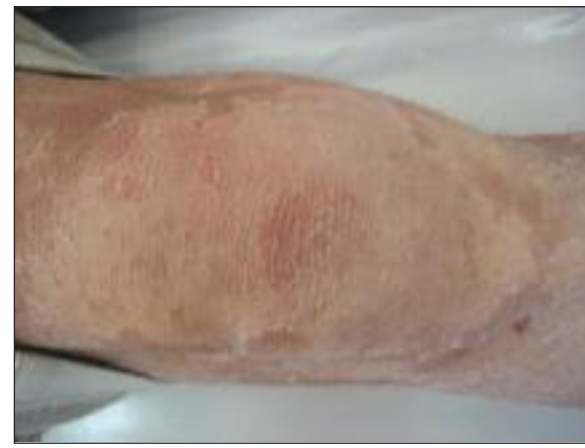

FIGURE 2: Figurate erythematous hyperkeratotic patches on knees.

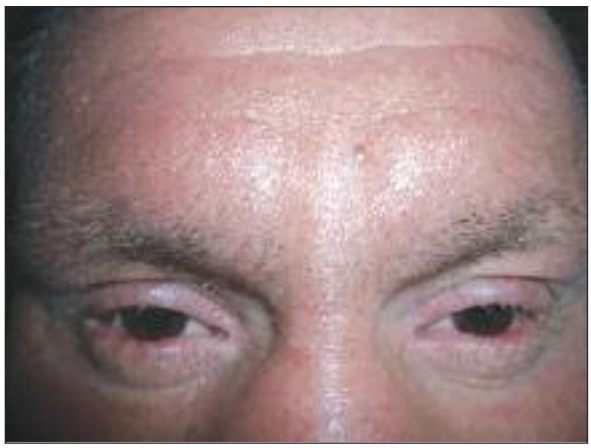

FIGURE 3: The eyebrows and eye lashes are sparse.

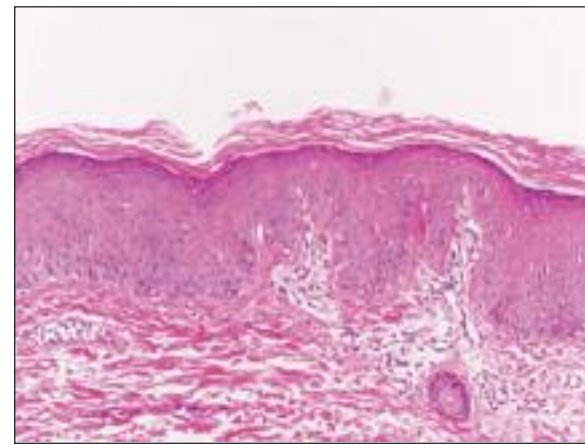

FIGURE 4: Hyperkeratosis, acanthosis and perivascular lymphocyte infiltrationin the upper dermis are seen (HE, x200).

ques which are especially localized to knees, elbows, dorsum of the hands and feet are found.[5] Diffuse palmoplantar hyperkeratosis is also characteristic. About half of the patients have sparse scalp hair and sparse or often absent eye brows and lashes.[3,5,6] Our patient developed figurate red-brown hyperkeratotic plaques since birth. These were localized over his body especially on knees, elbows. Bilateral plantar hyperkeratosis accompanied these plaques.

Chronic skin infections (bacterial, fungal) were noted in about half of the reported cases, although no consistent immunodeficiency has been demonstrated.[6] An essential component of KID syndrome is neurosensory deafness.[3,5-7] In only one case, hearing loss was reported to be conductory.[8] Hearing loss is usually detected in during infancy or early childhood, and develops by 7 years of age.[7] Caceros-Rios et al found deafness in all of their patients in their review.[9] Our case had bilateral neurosensory deafness since childhood.

A vascularizing keratitis of the corneas, the third major feature of KID syndrome occurs in about three-fourths of patients. The eye symptoms usually occur by early adolescence though at some times these may appear during the fourth decade.[3,10] In a recent review of 61 patients with KID syndrome, only 18 developed eye symptoms before 15 and it appears that some patients will never develop keratitis.[9] McGrae reported a patient with KID syndrome who developed keratitis at the age 22.[11] In our patient, the eye symptoms began at ten years old and were noted to be mild. He did not have keratitis, but had blepharitis and sensitivity to sun.

The unique cutaneous findings in KID syndrome begin in infancy. Hearing loss and keratitis complete this syndrome. Although deafness usually occurs at the age of 7, eye symptoms may appear later than expected, as it is seen in our case. That is why, the diagnosis of KID syndrome should not be ignored.

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