

Kindler syndrome: clinical particularities about two cases reports in two siblings.

Elharrouni Alaoui A, Baybay H, Chaoui R, Douhi Z, Elloudi S, Mernissi FZ

Department of dermatology CHU Hassan II, FES ,Morocco.

*Corresponding Author: Elharrouni Alaoui A, Department of dermatology CHU Hassan II, FES, Morocco.

Received date: March 17, 2020; Accepted date: June 03, 2020; Published date: June 16, 2020

Citation: Elharrouni Alaoui A, Baybay H, Chaoui R, Douhi Z, Elloudi S, Mernissi FZ.(2020) Kindler syndrome: clinical particularities about two cases reports in two siblings. Journal of Dermatology and Dermatitiss.5 (2); Doi: 10.31579/2578-8949/67

Copyright: ©2020 Elharrouni Alaoui Aicha, This is an open-access article distributed under the terms of The Creative Commons. Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstracts :

The Kindler syndrome, the fourth major type of hereditary epidermolysis bullosa (HEB), is a rare autosomal recessive genodermatoses, characterized by trauma-induced blistering, cutaneous atrophy, and progressive poikiloderma, in association with mucosal inflammation. We report tow new sibling cases of this rare syndrome.

Keyword : Kindler syndrome ; hereditary epidermolysis bullosa

Introduction:

Kindler’s syndrome (KS) is a rare type of genetic skin condition belonging to the class of bullous poikiloderma. It is characterized by skin fragility and blistering at birth followed by development of marked photosensitivity and progressive poikilodermatous skin changes in later years. We report two new sibling cases of this rare syndrome.

Case report:

This condition was seen in two girls aged 4 and 8 years, born of a first-degree consanguineous marriage with Kindler syndrome of varying degrees of severity. There are presented to us with a history of formation of multiple blisters, predominantly over the acral sites since birth. The one of patient also complained of photosensitivity.

Dermatological examination the older sister revealed a well-defined hemorrhagic flaccid bulla over dorsolateral aspect of the foot. Multiple hypopigmented scars with cigarette paper wrinkling were present over the dorsa of feet, hands, and elbows .there also had poikilodermia of gradual onset makes hypopigmented and hyperpigmented macules with telangiectasia, and atrophic scars over the face with angular cheilitis and gingival hypertrophy (figure :a-b-e).



And Cutaneous examination in younger sister revealed multiple hypopigmented and a few hyperpigmented macules of variable sizes, distributed over his face, neck, trunk, and limbs .Poikilodermatous skin changes were present. The overall texture of the skin was xerotic with marked cutaneous atrophy. The palms showed hyperkeratosis with diminution of palmar creases. Skin over the hands and neck was dry, atrophic and photosensitive to the sunlight. The patient was short stature (figure: c-d). Clinical diagnosis and genetic study were in favor of Kindler syndrome



FIGURE 1(a_b_c_d_e): Clinical features of Kindler syndrome in two girls aged 4 and 8 years. Poikiloderma involving the neck and trunk, skin atrophy, and gingival hypertrophy.

Discussion:

Kindler syndrome (KS) is a rare autosomal recessive genodermatosis, characterized by skin fragility and blistering at birth followed by development of marked photosensitivity and progressive poikilodermatous skin changes in later years(1). KS is associated with mutations in the KIND1 (FERMT- 1) gene that result in either hypomorphic, or complete loss of Kindlin-1 activity(1). Clinical findings in KS are increased skin fragility, acral blistering, photosensitivity, atrophy, and poikiloderma. The photosensitivity usually decreases over time, coinciding with decreased blister formation by 10-12 years of age, although some degree of photosensitivity usually persists even after that age (1-2). Poikiloderma, and atrophy generally occur at sun-exposed sites, but it can also present at non-exposed sites. palmoplantar keratoderma, pseudosyndactyly, leukokeratosis of lips and oral mucosa, pseudoainhum, xerostomia, phimosis, dental caries, periodontitis, Gingivitis and periodontitis are also prominent features(2-3). Gastrointestinal symptoms, including constipation and severe colitis, can also occur in KS. Diagnosis of Kindler syndrome is based upon clinical evidence. Electron microscopic examination is used in particular to rule out congenital bullous epidermolysis(4). Detachment of layers at two or three different levels in relation to the dermal-epidermal junction described in the literature forms a specific but inconsistent feature of Kindler syndrome(2). The management of KS is largely symptomatic, encompasses protective measures that include sun protection and care of acral bullae and wounds by appropriate topical and systemic antibiotic treatment (2-5).

Conclusion:

Kindler syndrome is a rare syndrome. Its diagnosis is practically clinical. Different levels of cleavage have been described in electron microscopy. We have reported two new observations of two sisters who borned as a first-degree consanguineous marriage in a family whose clinic and genetic study were the keys to diagnosis.

Reference:

1. Leila Youssefian et al. The Kindler Syndrome: A Spectrum of FERMT1 Mutations in Iranian Families. Journal of Investigative Dermatology accepted article preview 19 January 2015.
2. Suman, et al. Kindler's syndrome: A rare case report. Departments of Prosthodontics, and 1Periodontics, SGRD Institute of Dental Sciences and Research, Amritsar, Punjab, India .Contemporary Clinical Dentistry | Apr-Jun 2014
3. Nadia El Fekih et al. Kindler syndrome: clinical and ultra-structural particularities, a propos of three cases. 0242-6498
4. Elizabeth K Duperret & Todd W Ridky (2014) Kindler syndrome in mice and men, Cancer Biology & Therapy, 15:9, 1113-1116.
5. Schumann H. Epidermolysis bullosa. An update. Hautarzt 2009;60:614—621.