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Papillon Lefevre Syndrome-An Atypical Late Onset Presentation

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ABSTRACT

Papillon-Lefevre Syndrome (PLS) is a rare autosomal disorder characterized by palmar plantar keratosis and aggressive periodontitis. Herewith, we report the first case of PLS to our knowledge presenting with an atypical late onset of periodontal disease component from the MENA region.

Keywords: Papillon-Lefevre Syndrome, autosomal disorder, palmar plantar keratosis, periodontal disease, MENA region

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1. Introduction

Palmoplantar keratodermas (PPKs) is a heterogeneous group of disorders characterized by thickening or hyperkeratosis of the palmar and plantar skin with or without other associated clinical features. This thickening and hyperkeratosis of the palmar and plantar skin, can be hereditary or acquired; diffuse, focal, or punctuate; and transgrediens (defined as contiguous extension of hyperkeratosis

beyond the palmar and/or plantar skin-transgrediens behavior indicates spread of the keratoderma to involve the dorsa of the hands and feet) or progrediens (progrediens behavior signifies disease progression with age). PKK's can also be distinguished from each other on the basis of histopathologic findings (the presence or absence of epidermolysis by the presence of other associated abnormalities, and the pattern of inheritance. PPKs are further distinguished by their mode of inheritance and by the presence of certain associated clinical features.[1,2]

The Papillon-Lefevre Syndrome (PLS) is a type of PKK. It was first described by two French dermatologists Papillon and Lefévre in 1924 as "Mal de Meleda". The cardinal features of PLS are palmar-plantar hyperkeratosis and premature loss of deciduous and permanent teeth due to aggressive periodontitis.[1] PLS (PLS; OMIM 245000) is a rare autosomal recessive disorder and is caused by mutations in the cathepsin C (CTSC) gene on chromosome 11q14 and involves the skin, gingiva, and teeth.[3] It has a frequency of 1–4 persons per million with no sex predilection.[4]

PLS is also associated with, intense gingivitis as early as infancy with concomitant alveolar bone loss resulting in loss of deciduous teeth. PLS affected individuals show enhanced susceptibility to infections (furunculosis, skin abscesses, hidradenitis suppurativa). PLS is also frequently associated with anodontia/oligodontia, dysplastic/thick/grooved fingernails, hamartoma/tumors of the mouth, teeth anomalies, arachnodactyly, intracranial calcifications and thin/hypoplastic/hyper convex fingernails. Decreased body hair, increased body hair, skin tumors/lumps, malodorous hyperhidrosis, sparse/absent scalp hair (generalized) are occasionally seen with PLS.[5] Anomalies of chemotaxis and phagocytosis by polymorphonuclear leukocytes have been observed.[5,6,7]

PLS affected individuals loose their complete permanent dentition barring the third molars between 14-17 years leading to a toothless period which is followed by the eruption of third molars. The literature search in PubMed revealed only a few family case series with a late presentation or partial expression of either its dermal lesions or the periodontal presentation.[8,9,10] With this, we report the first case to the best of our knowledge from the Middle East North Africa region with such an atypical presentation.

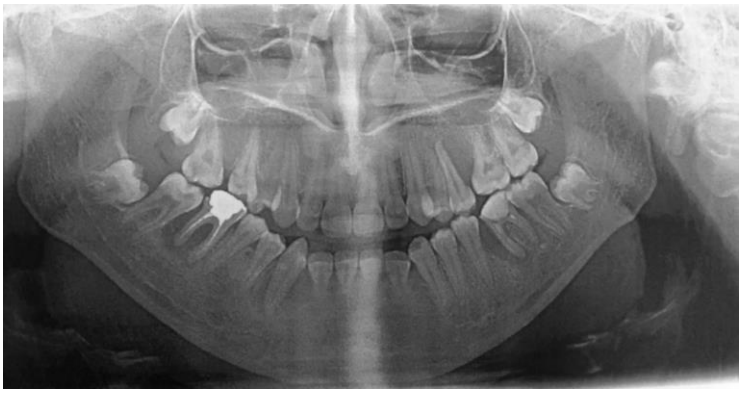
2. Case Presentation

A 16-year-old female reported to the outpatient dental clinic complaining of swelling and redness in lower front gums. The otherwise noncontributory medical history revealed skin thickening and scaling on the palms and soles since birth. Family history revealed consanguineous marriage of non-symptomatic healthy parents with all the siblings showing similar skin lesions since birth (3 sisters and 2 brothers). Her brothers had no periodontal lesions while they had skin lesions of PLS. One of the patients elder sisters had lost all her teeth. Her paternal uncle had a similar history. Unfortunately, the other family members did not consent to publication of clinical photographs. The presented subject did not report the early loss of her primary dentition. Extraoral examination revealed well-demarcated, yellowish, hyperkeratotic plaques bilaterally seen over the skin of her palms and soles extending onto the dorsal surfaces. These skin plaques were non-tender and increased with water exposure. Hypohydrosis of skin with dystrophy,

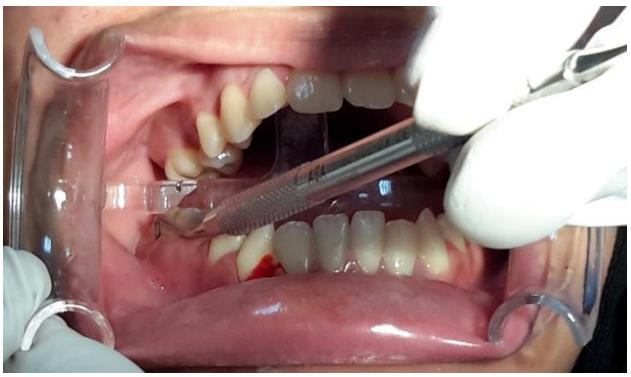
transverse grooving and malformation of nails was noted.

Intraoral examination revealed hyperemic oedematous gingiva in the mandibular canine region. She had minimal calculus and slight subgingival plaque. Careful periodontal probing revealed multiple deep periodontal pockets and discomfort while eating. The Radiographic examination (orthopantomogram) showed the presence of generalized destruction of the alveolar bone nevertheless a "floating in the air" appearance was not seen. Routine hematological examination produced results within normal limits. Thus correlating the family history and clinical presentation of the presence of marked palmar-plantar hypertrophy with a relatively late-onset destructive periodontitis, it is diagnosed as Papillon-Lefevre syndrome.

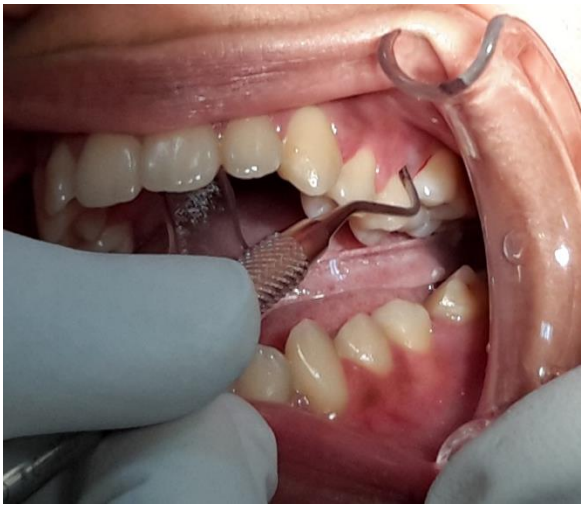
The treatment was planned with oral hygiene instructions, scaling and root planing with adjunctive systemic antibiotics (amoxicillin 500mg tid+ Metronidazole 200mg tid for 7 days). The patient was explained that periodontally unsalvageable teeth have to undergo extraction following the initial evaluation. The patient has also been referred to a dermatologist for treatment of skin lesions.



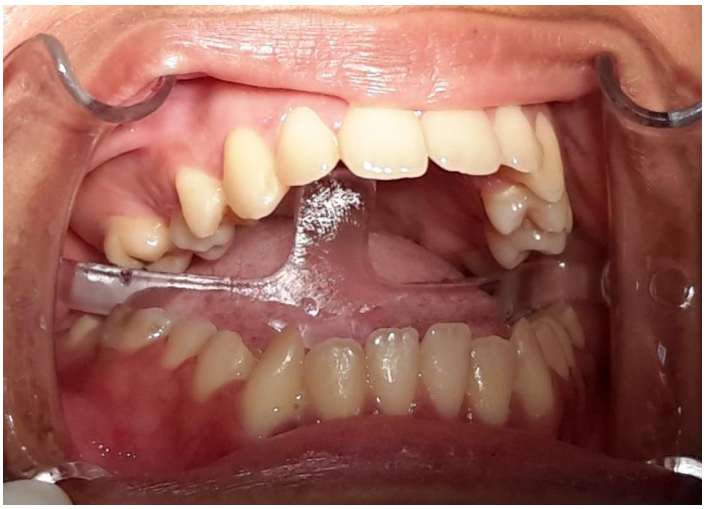
Fugure: 1 OPG showing bone loss in multiple teeth



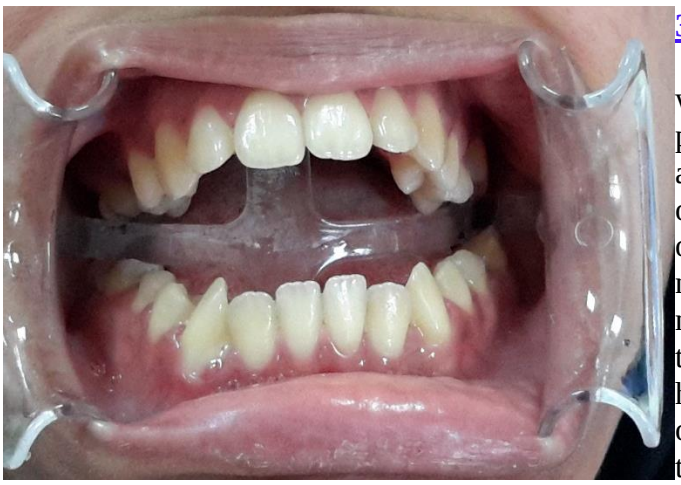
Fugure:2 Shows deep periodontal pocket and inflammed area in relation to 33



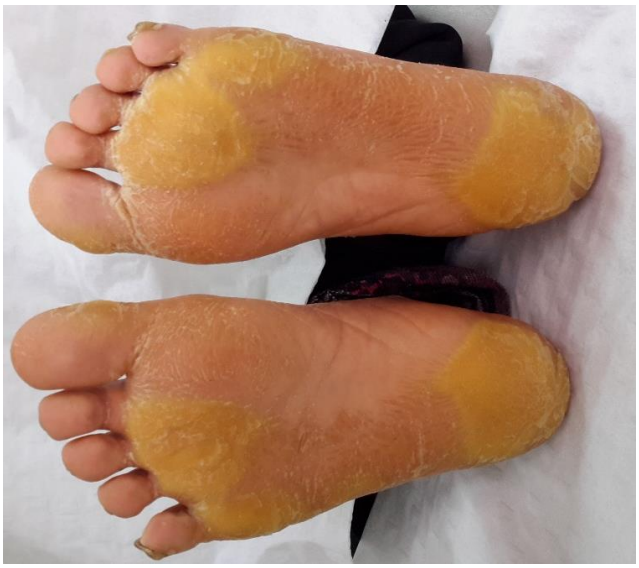
Fugure:3 Shows deep periodontal pocket.



Fugure:4 Intra Oral view.



Fugure:5 Intra Oral view.



Fugure:6 Plantar Keratosis

Fugure:7 Palmar keratosis spreading to the dorsal side.

3. Discussion

PLS is a rare entity characterized with palmar-plantar keratosis and aggressive periodontitis. In our patients, we were not able to perform any genetic analysis and the other patients who were the other members of the family except the presented one did not consent for photographs. The family members had different presentations. With the case presented with clinical photographs had palmar plantar keratosis and relative late onset of aggressive periodontitis. Various treatment options have been reported in literature, which include professional cleaning, administration of antibiotics, administration of oral retinoids, extraction of the affected teeth and replacement with either removable prosthetics or implants.[11,12,13] A conservative approach was taken by us in treating the patients a combination of professional debridement of the periodontal lesions with an oral administration of antibiotics in patients who showed up clinical features of periodontitis. The sibling who had lost all the teeth was administered denture.

To conclude, Herewith we present the first known case with an atypical presentation of PLS with late onset of aggressive periodontitis from a family with PLS in MENA region.

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