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CASE REPORT

Oral Manifestation of Papillon-Lefevre in Two Siblings

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ABSTRACT

Background: *Papillon-Lefèvre Syndrome (PLS) is a very rare genetic disorder (autosomal recessive) that usually affects infants between the ages of one and five years. PLS is characterized by palmar-plantar hyperkeratosis and premature loss of primary and permanent teeth with extensive, severe, and rapid destruction of the alveolar bone, leading to the premature loss of both teeth.* ***Objective:*** *To report on the management of PLS patients.* ***Case:*** *A 9-year-old girl and a 2.5-year-old boy from Nganjukto General Hospital, Pediatric Dentistry Division, RSGM Airlangga University with the main complaints of upper left tooth pain, rocking, difficulty eating, erythematous hyperkeratosis hands and feet, periodontal inflammation, mobility. bad teeth.* ***Case Management:*** *Nine-year-old girl: Teeth #26 and #46 could not be retained and extracted due to buccoversion and severe mobility. Periodontal supportive therapy was periodontal debridement, scaling and root planing and oral hygiene counseling with 0.20% chlorhexidine gluconate. Two-and-a-half-year-old boy: Too young, he is observed every 3 months. It is recommended to take vitamin B complex in appropriate doses and fruit juice as an additional supplement.* ***Conclusion:*** *Intensive periodontal treatment and administration of antibiotics only delays the progression of periodontal disease and cannot be used to prevent primary and permanent teeth. Extensive and repeated prosthodontic treatment is required to provide functional teeth to children during their period of jaw growth.*

Keywords: *Aggressive Periodontitis, Papillon-Lefèvre Syndrome, Palmar-plantar Hyperkeratosis, Siblings*

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INTRODUCTION

In 1924, Papillon-Lefèvre Syndrome (PLS) was discovered by doctors Papillon and Lefèvre from France.[1,2,3] The characteristic feature of PLS is diffuse or localized hyperkeratosis on the palms and soles of the feet (palmar-plantar hyperkeratosis).These signs are always associated with generalized aggressive periodontitis, and are recognized by the presence of severe inflammation and degeneration of the structures surrounding and supporting the teeth (periodontium). [4,5,6]

Papillon-Lefèvre Syndrome (PLS) is a rare autosomal recessive disorder. The syndrome is believed to affect one to four individuals per million.[1] An increased prevalence was observed in the offspring of relatives; Kinship between parents was documented in one third of the cases studied. The predominance of race in the case of Papillon-Lefèvre Syndrome is absent and for the sexes males and females are equally affected in this regard. Susceptibility to infection can increase in about 20% of cases in individuals diagnosed with PLS, but studies have not found that show a decrease in life expectancy.[7]

Primary teeth in children often experience loose and fall out. However, if the permanent teeth are not treated, they will decay and even fall out.[7,8] Other signs and symptoms of this syndrome include frequent pyogenic skin infections, nail dystrophy, and hyperhidrosis. Parents with very close ancestry are shown in between 20% and 40% of cases. Functional or quantitative neutrophil abnormalities, and 50% are immunocompromised are usually associated with patients diagnosed with PLS. Patients with PLS appear to have a particular predisposition to develop pyogenic liver abscesses. Pyogenic liver abscess is a rare presentation.[4,9]

A recently discovered genetic abnormality in PLS has been mapped to chromosome 11q14-q21 involving a cathepsin C mutation. Studies show more than 90% of PLS patients have reduced cathepsin C activity.[3,10,11] Impaired chemotic and phagocytic functions of polymorphonuclear leukocytes (PMNs) have been described in many reports. In contrast to the above studies, however, PMN chemotaxis reported normal. Several reports also discuss lymphocyte function in PLS patients.[12]

The effects on the periodontium appear immediately after tooth eruption, as the gingiva becomes erythematous and edematous.[5] Plaque accumulation in deep fissures and halitosis may occur. The primary incisors are the most affected teeth first and can show marked mobility, especially by the age of 3 years. By 4 or 5 years of age, all of the primary teeth may have changed. Most teeth fall out at the age of 14-15 years. Damage to the alveolar bone often results in jaw atrophy. In such cases the patient is often edentulous at an early age. This case report presents a brief overview of two siblings of a patient diagnosed with Papillon-Lefèvre syndrome and describes the clinical presentation in a case with characteristic dental and dermatological findings. Symptoms experienced by PLS patients are psychological, social, and aesthetic symptoms. Management has its own challenges and handling PLS cases requires a multidisciplinary approach from dental surgeons, dermatologists and pediatricians.

CASE

There were two patients from General Hospital of Nganjukto Pediatric Dentistry Department, Universitas Airlangga Dental Hospital. Nine years old girl and two and half-years old boy with chief complaint of tooth ache on the upper left and loose, also difficulty in eating. No family members, other relatives, or even the patient's parents showed similar manifestations. The skin lesions started to appear after two and half years of age. There was no history of other serious illness or susceptibility to infection in areas other than the oral cavity.

Dental history revealed eruption of primary teeth in normal chronological order. The girl was around three years old; the gingiva was

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red, swollen, and painful. Furthermore, the teeth become mobile and exfoliated. The girl was edentulous by the age of three and half years. Intraoral examination showed generalized severe periodontitis manifested by redness of gingiva, generalized inflammatory gingival enlargement. Deep periodontal pockets (>6 mm) were present around all permanent teeth. There are moving, drifting (buccoversion), and extruded teeth. The patient has difficulty chewing and severe halitosis. Tooth size in normal shape and size (figure 1,2).

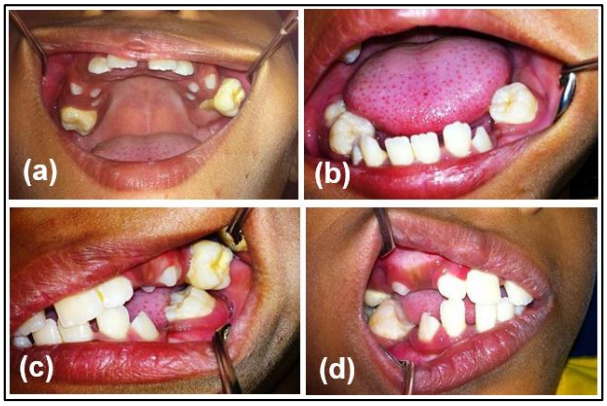

Figure 1. Intraoral examination with severe gingival inflammation in a girl. a) Pre-operative examination of the upper jaw. b) Pre-operative examination of the lower jaw. c) Occlusion in the left lateral view. d) Occlusion in right lateral view.

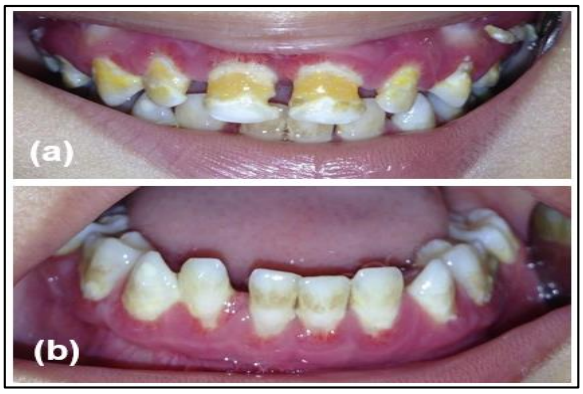

Figure 2. Intraoral examination with severe gingival inflammation in a boy. a) Pre-operative examination of the upper jaw. b) Pre-operative examination of the lower jaw.

Dermatological examination showed hyperkeratosis of palms and soles in the form of well- demarcated plaques. The hyperkeratosis was exacerbated during the winter months (figure 3,4). Radiographic examination revealed a severe alveolar bone loss of all erupted teeth with less than one third of alveolar bone remaining giving the teeth a “floating in air” (figure 5).

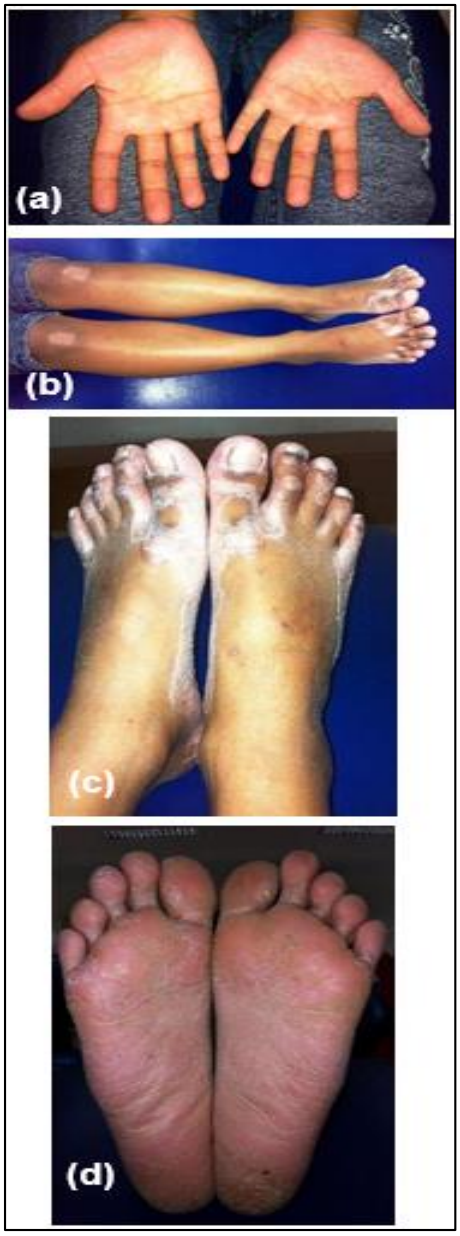

Figure 3. Palmar-Plantar Hyperkeratosis in a girl. a)Palmar surface of Hand. b)Front view of Leg. c)Dorsal surface of Foot. d) Plantar surface of Foot.

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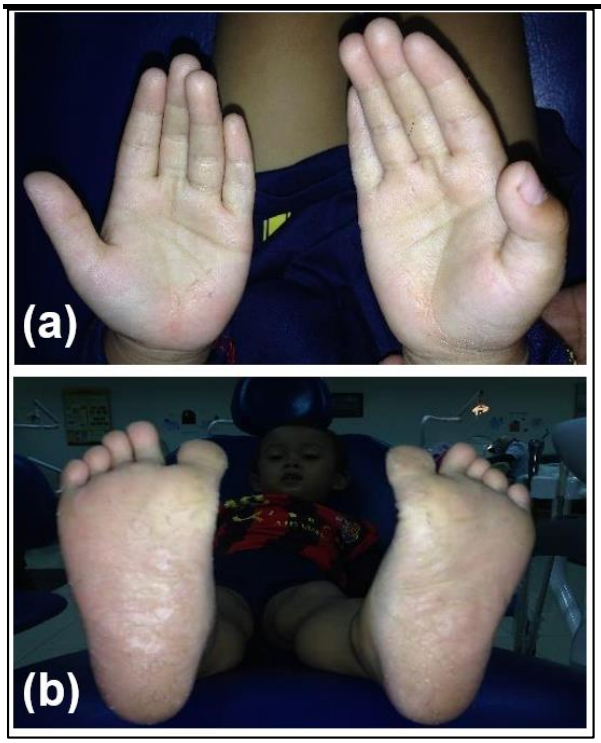

Figure 4. Palmar-Plantar Hyperkeratosis in a boy. a)Palmar surface. b) Plantar surface.

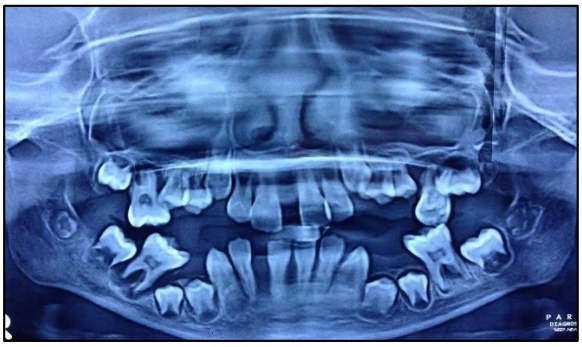

Figure 5. Radiographic Examination

CASE MANAGEMENT

In the history of the two siblings there was no systemic problem, but palmar-plantar hyperkeratosis was observed. All the primary dentition molars were lost early in both upper and lower jaw (figure 6). In addition to third degree mobility, alveolar bone loss, severe gingivitis and plaques, in the light of these findings Papillon-Lefèvre Syndrome was diagnosed in both patients. First, the two siblings,9-year-old girl and a 2.5-year-old was prescribed an antibiotic of amoxicillin (250 mg, 2x1, one week) and metronidazole (250 mg, 2x1, one week) and a mouth rinse of 0,2% chlorhexidine gluconate (2x1, one week), and educated for oral hygiene. Vitamin C and B complex is further prescribed. Her mother gave some fruit juice at home, such as tomato, avocado, orange, etc.

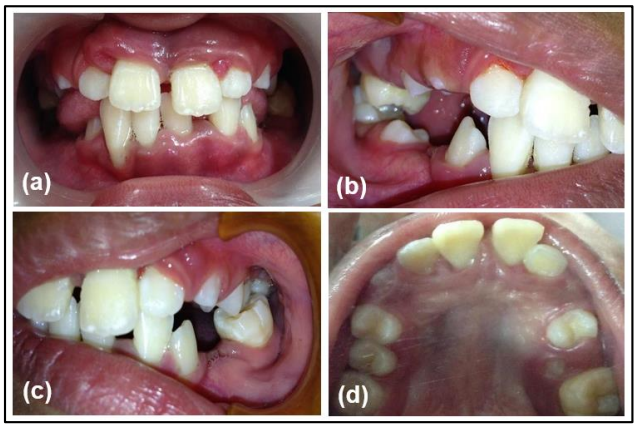

Figure 6. Intraoral Examination Observation. a) Occlusion in front view. b) Occlusion in right lateral view.c) Occlusion in left lateral view. d) Upper jaw view.

Another treatment for the palmar-plantar hyperkeratosis to minimize the effect and appearance of scales on the skin of the patient, suggested using Kloderma or olive oil after taking a bath, based on consultation with Dermatologist (Figure 7). The radiographic examination after #26 and #46 were extracted because of its buccoversion and severe mobility (Figure 8). The boy was too young, so he was observed every 3 months. Panoramic for the boy is unavailable because of cooperative issues.

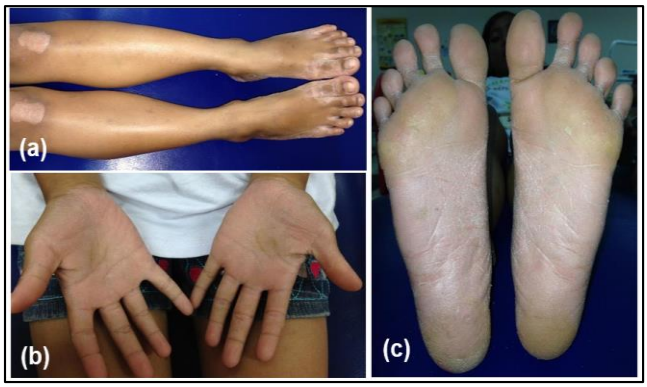

Figure 7. Palmar-Plantar Hyperkeratosis after Treatment. a) Front view of Leg. b) Palmar surface. c) Plantar surface.

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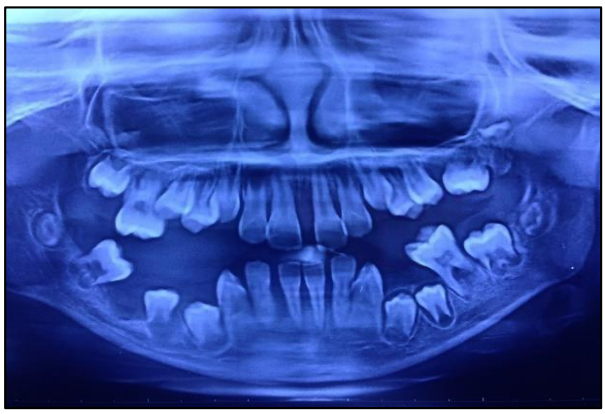

Figure 8. Radiographic after five months.

DISCUSSION

Papillon-Lefèvre syndrome is an inherited autosomal recessive disorder. There is no influence of genes and a family history of disease that can cause PLS. Increased prevalence occurs if there is blood relationship with parents, but is not influenced by gender or race. [13,14,15]

Palmar-plantar hyperkeratosis and rapidly progressive generalized periodontitis accompanied by severe alveolar bone destruction are clinical signs of PLS. This causes premature loss of primary and permanent teeth. [16,17,18]

High levels of Aggregatibacter actinomycetemcomitans are directly related to the etiology and pathogenesis of PLS periodontitis so that successful treatment plans with specifically working antibiotics are the main treatment.[16] Treatment of Papillon-Lefèvre Syndrome is the removal of all primary teeth and replacing them with complete dentures.[4] The effectiveness of extraction with different antibiotics will prevent or delay permanent tooth loss if combined with periodontal treatment.[19] Studies have shown that *Aggregatibakcter actinomycetemcomitans* was effective with amoxicillin/metronidazole and may be included in the therapeutic protocol of individual with Papillon-Lefèvre Syndrome.[3] Although there is a clear association between the presence of *Aggregatibacter actinomycetemcomitans* and the Papillon-Lefèvre Syndrome, there is no evidence of cause and effect. The majority of PLS patients have lost their teeth by the age of 13, despite different treatments.[20]

Extraction therapy with antibiotics accompanied by periodontal treatment can be intervened in individuals who have been identified as having PLS before the eruption of permanent teeth. Choosing the right treatment is not so easy when the patient seeks dental treatment at the mixed dentition stage or later.[16]

Progressing periodontal disorder main to tooth loss is a first-rate trauma in this patient. Extensive and repeated prosthodontic treatment may additionally become necessary to offer the kids with a functional dentition at some stage in the growth length in their jaws. Edentulousness and location of complete dentures that want to be renewed at quick intervals is a similarly unappealing option. Suggestions for further examination are to always monitor the balance of occlusion, balance the strength of mastication, and take vitamins to boost the immune system. In this case, a toxicological method is needed to determine the concentration of heavy metals in human hair and nails, such as Hg, Cu, Ni, Pb, Co, and Cd. Extensive and repeated prosthodontic treatment is required to provide functional teeth to children during their period of jaw growth.

CONCLUSION

The combination of intensive periodontal treatment and administration of antibiotics only temporarily delays the progression of periodontal disease and cannot be used as a means of preventing the loss of primary and permanent teeth. The results of this case report indicate that the management of periodontitis in PLS patients becomes more complicated when the initial periodontitis occurs in the mixed dentition. In this situation, control of the progression of periodontal decay can be accomplished and the minimization of tooth loss is greatly improved.

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