

A Case Report: Papillon Lefevre Syndrome

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Submitted: 05 Oct 2019; Accepted: 12 Oct 2019; Published: 26 Oct 2019

Abstract

Study Background: Papillon Lefevre Syndrome (PLS) is an autosomal recessive inherited genetic disorder characterized by palmoplantar hyperkeratosis and premature loss of deciduous and permanent teeth. Mutation of cathepsin C gene has been detected in the patient with PLS. In this case report, oral and radiographic findings of one PLS patient were discussed. 14 year old male patient diagnosed with PLS after radiographic, dermatologic and dental examination was referred to our clinic due to severe gingival inflammation and mobility. Severe gingival inflammation, alveolar bone resorption and multiple teeth loss due to periodontal reasons were observed. Mild palmar and moderate plantar hyperkeratosis were observed but no other systemic problems were detected.

Methods: Non-surgical periodontal treatment was performed among multiple sessions and in each session subgingival tissues had been irrigated with %2 chlorhexidine irrigation solution. To reduce severe gingival inflammation antibiotherapy has been prescribed. Hopeless teeth were extracted.

Results: After non-surgical periodontal treatment and antibiotherapy severe gingival inflammation was reduced but not eliminated completely, mobility score of some teeth reached to zero score, patient was motivated about oral hygiene and was taken into maintenance phases. After having extracted hopeless teeth patient was directed for prosthodontic rehabilitation.

Conclusion: PLS is a rare autosomal recessive disorder. With PLS-specific dermatologic findings and characteristic periodontal view, it is possible to be able to identify the disease in its very early stages. Early diagnosis of the disease and institution of an appropriate periodontal and antimicrobial treatment might improve the prognosis.

Introduction

Papillon Lefevre syndrome (PLS) was first described by two French physicians, Papillon and Lefevre, in France in 1924 [1]. It is an autosomal recessive inherited genetic disorder with the incidence one to four per million with no sex and racial predominance [2]. High incidence of PLS is seen in consanguineous marriages [3]. This disease is characterized by redness and thickening of the soles and palms, severe destruction of periodontal tissues affecting both deciduous and permanent teeth. It is caused by mutation located on long arm of chromosome 11, which involves mutations of the cathepsin C gene [4]. Dermatological, immunological, hematological, oral and microbial aspects are seen. Other symptoms include hyperhidrosis, arachnodactyly, intracranial calcification, increased susceptibility to infections, and mental retardation [5]. Deficiency of cathepsin C function will result in loss of immunological response, leading to liability of infection. Impairment of natural killer cell cytotoxic function is the first consistent immune dysfunction in PLS. This suggests that the impaired natural killer cell cytotoxicity might contribute to the pathogenesis of PLS-associated periodontitis [6].

Materials and Methods

14 year old male patient diagnosed with PLS was referred to our clinic due to severe gingival inflammation and mobility. After radiographic, dermatologic and dental examination diagnosis had been confirmed. Ortopantomography and periapical radiography was used for radiographic examination and alveolar bone resorption was observed. Nothing unusual was seen in hand-wrist radiography.

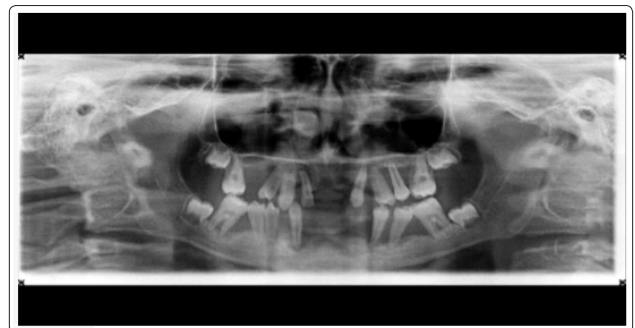


Figure 1: Ortopantomography



Figure 2: Periapical radiography



Figure 3 and 4: Hand-wrist radiography

Patient's family included consanguineous marriages and his pedigree was obtained.

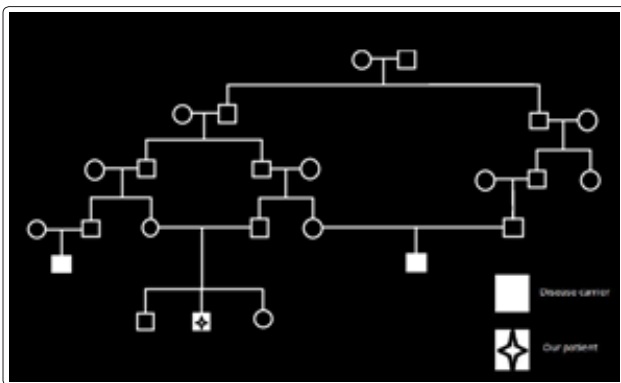


Figure 5: Pedigree

In intraoral examination severe gingival inflammation and mobility, spontaneous bleeding, deep probing depths, poor oral hygiene and absence of multiple teeth were observed. Patient was suffering from eating and talking dysfunctions. Intraoral examination datas had been documented through periodontal chart created by University of Bern, Department of Periodontology.



Figure 6: Intraoral photography of first examination

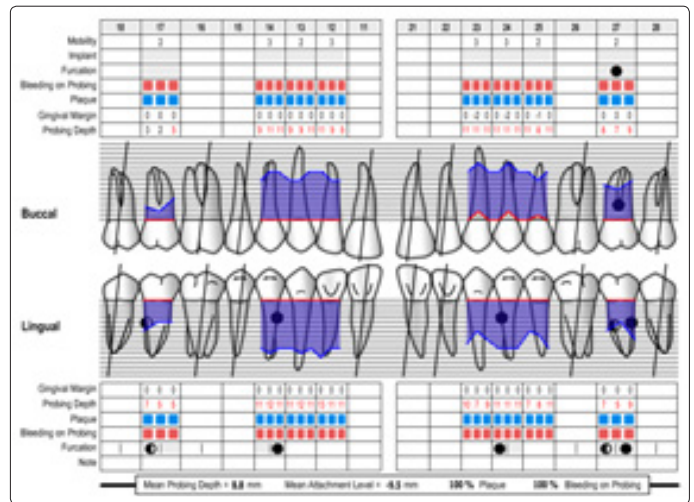


Figure 7: Datas of upper jaw at first intraoral examination

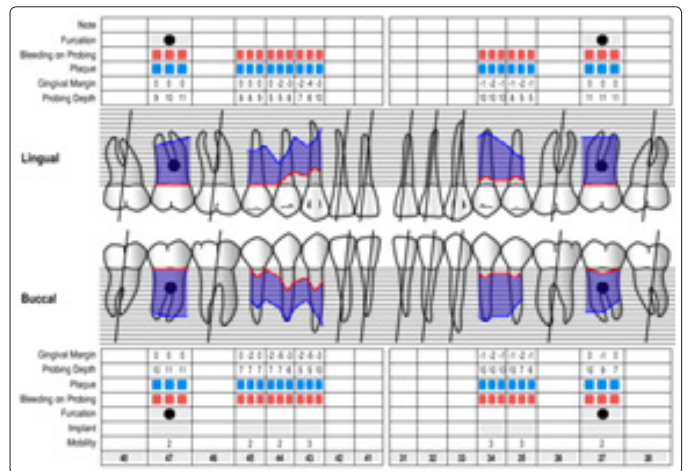


Figure 8: Datas of lower jaw at first intraoral examination

In dermatological examination hyperkeratozis was seen over palms and soles but no other systemic problems were detected.



Figure 9, 10 and 11: Palmoplantar hyperkeratosis

Scaling and root planing (ScRp) treatments had been applied repeatedly once a week. Right after first ScRp treatment, with consideration of patient's weight and medical condition, 625 mg of amoxicillin and clavulanic acid combination had been prescribed orally twice a day for 7 days and 250 mg of metronidazole three

times a day for 7 days. Subgingival tissues had been irrigated with %2 chlorhexidine gluconate solution following ScRp treatments and %0.15 chlorhexidine gluconate mouthwash had been recommended twice a day. Patient had been trained about oral hygiene. After non-surgical periodontal and antimicrobial treatment severe gingival inflammation was reduced but not eliminated completely, mobility score of some teeth reached to zero score, patient was motivated about oral hygiene and was taken into maintenance phases.



Figure 12: Intraoral photography of post-treatment examination

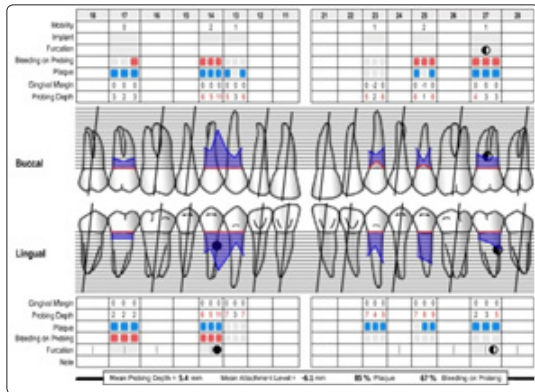


Figure 13: Datas of upper jaw at last intraoral examination

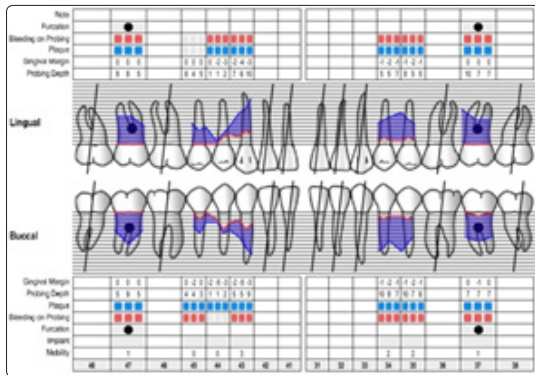


Figure 14: Datas of lower jaw at last intraoral examination

Two hopeless teeth were extracted.



Figure 15: Intraoral photography of post-extraction

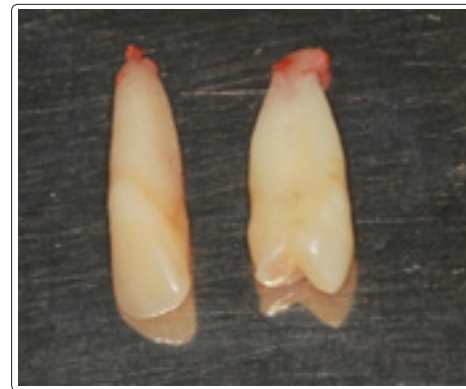
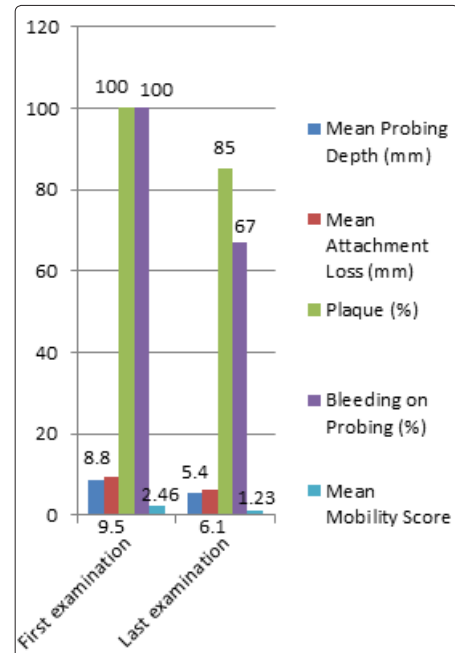


Figure 16: Extracted teeth

Results

After non-surgical periodontal and antimicrobial treatment PLS-associated periodontitis had been taken under control partially and patient was motivated about oral hygiene. After having extracted hopeless teeth patient was directed for prosthodontic rehabilitation.

Table 1: Comparison of examination datas before and after treatment



Discussion

PLS is a rare autosomal recessive disorder. With PLS-specific dermatologic findings and characteristic periodontal view, it is possible to be able to identify the disease in its very early stages. Early diagnosis of the disease and institution of an appropriate periodontal and antimicrobial treatment might improve the prognosis.

References

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