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CASE REPORT

FOLLOW UP FOR PEDIATRIC DENTAL PATIENT WITH PAPILLON-LEFEVRE SYNDROME, IN MONGOLIA

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Background: Papillon-Lefevre Syndrome (PLS) is a rare genetic disorder, characterized by palmoplantar keratosis, aggressive periodontitis and premature edentulous primary and permanent dentition at a very young age.
Case description: A 3.6-year-old male patient reported to the central dental hospital, with a complaint of tongue ulcer and exposed root of deciduous teeth. Intraoral examination of the patient had shown the absence of central incisors in maxilla and in the mandible presence of first primary premolars and right primary canine. The anterior third of the tongue was a traumatic ulcer in the maxillary left canine and lateral incisor
Practical implications: We have followed up patient with PLS and the traumatic tongue ulcer of patient with immunodeficiency was recovered. Dentists are very important specialists for the diagnosis and management of PLS.

***Keywords*:** Edentulous, alveolar bone resorption, genetic disorder, immunoglobulin A, tongue ulcer

INTRODUCTION

Papillon-Lefèvre syndrome (PLS) was initially described by French physicians Papillon and Lefevre in 1924, with palmoplantar hyperkeratosis accompanied by early onset periodontitis and premature loss of primary as well as permanent teeth [1,2]. PLS is a rare autosomal recessive disorder featured by destructive periodontitis beginning in childhood, diffuse, transgradient palmoplantar keratoderma, premature loss of permanent teeth, and frequent cutaneous and systemic pyogenic infections [3-5].

In Mongolia, as of 2017-2021, the prevalence of congenital anomalies is 8.6 per 1000 live births and the common types were congenital malformations: circulatory system (24.8%), musculoskeletal system (16.7%), multiple organ defect (14.1%), cleft lip and palate (13.8%), and the digestive system (7.7%). Congenital anomalies were statistically significantly higher among male, low birth weight infants and parents aged 35 or over [6].

Indeed, various pathologies and syndromes can affect periodontal health in different ways: systemic conditions

(such as PLS), pre existing conditions (patients with uncontrolled diabetes), local factors and inflammatory conditions [4,5]. This article describes a clinical presentation of a boy diagnosed with PLS in the dental clinic.

CASE PRESENTATION

A 3.6-year-old male patient reported to the Central dental hospital, with a complaint of tongue ulcer and exposed root of deciduous teeth.

According to the patient's parents, his deciduous teeth erupted normally, but exfoliated at the age of 3. He had suffered congenital immune deficiency and sensory neuropathy. Pyogenic infections were detected 4-5 times in different areas of his body, such as index finger nail, the right femoral bone, the left radius and the various parts of the skin.

On physical examination, he was skinny and weak, hobbled walking and held onto his mother occasionally, closed narrow eyes, hand finger not stretched out completely, abnormal fingernails, and hyperkeratotic lesions on the palm were observed. And he has a plaster cast in his left hand between his wrist and middle arm.

His father has a cleft palate and lip (Fig. 1) and #15, #25, #37 teeth have a shortened root in his mother's orthopantomograph (Fig. 2). There were no similar complications in his family history.

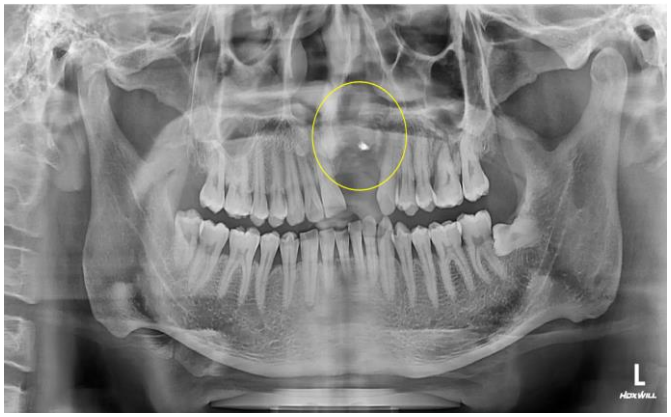

Fig. 1. Panoramic radiograph of the patient's father (Yellow circle located in the cleft palate, there is no bone tissue between nasal and oral cavities. The opaque shadow may be a residual dental biomaterial from anamnesis patients father)

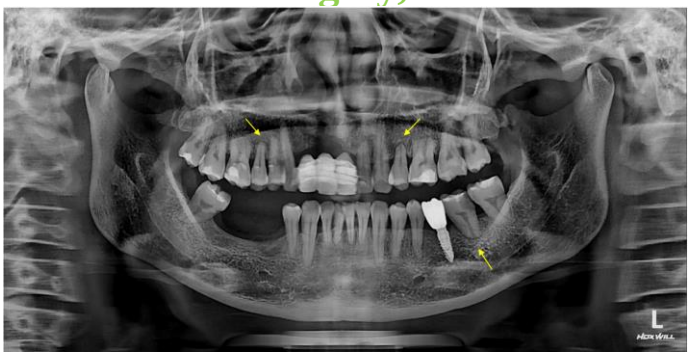

Fig. 2. Panoramic radiograph of the patient's mother (The yellow arrows indicate the shortened root of maxillary second premolars and mandibular left second molar)

The initial oral examination of the patient had shown the absence of primary maxillary central incisors and presence of maxillary molars, canine and the lateral incisors (Fig. 3A). In the mandible presence of first primary premolars and right primary canine. And also other mandibular primary teeth were missing (Fig. 3B). A traumatic ulcer was present on the anterior third of the tongue, caused by contact with the maxillary left canine and lateral incisor (Fig. 3C) [7].

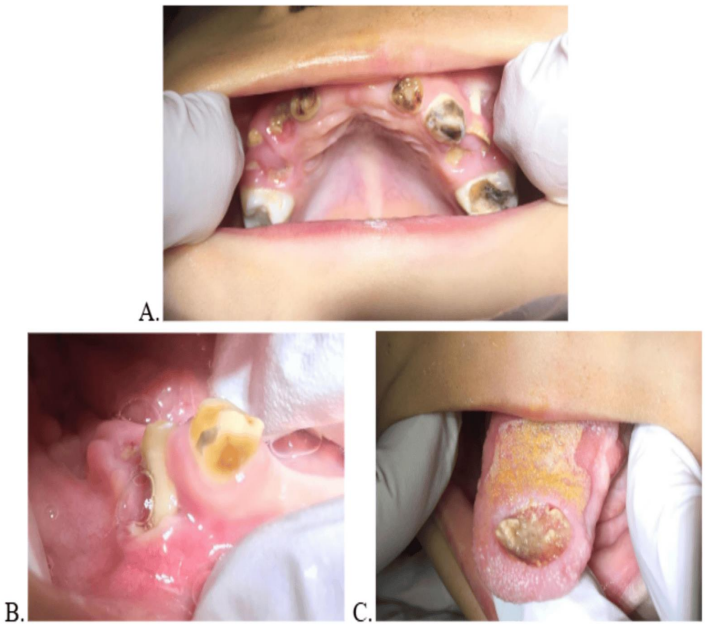

Fig. 3. Patient's oral photo; A-maxillary dental arch, B-lower left canine and molar; C-The tongue ulcer [7]

A first panoramic radiograph displayed the lower first permanent molars in the lower dental arch and all the permanent tooth germs, except #45 in the mandible bone, when he was 4.5 years old. In the maxilla there

were all permanent tooth germs, which are different stages of tooth development (Fig. 4A). The radiopaque image on the mesial side of the crown of the mandibular right first permanent molar and alveolar bone was local horizontal resorption around this tooth.

A second panoramic radiograph, when he was 5.9 years old, displayed the lower first permanent molars and central incisors in the lower dental arch and the all permanent tooth germs were at a more advanced stage than had being a 4.5-year-old. The new radiopaque image in the coronal occlusal side of the mandibular right first permanent molar and alveolar bone was restored. The first permanent molar in the upper dental arch and the tooth germs of the permanent premolar, canine and incisors were overlapped (Fig. 4B) [7].

From the next following panoramic radiograph, when he was 6.3 years old, we could see the central and lateral incisors in the mandibular dental arch; the central incisors and left first premolar were erupted in the maxillary dental arch. Therefore, the roots of the prematurely erupted permanent teeth developed normally. A new radiopaque image was observed on the coronal-occlusal side of the mandibular left first permanent molar (Fig. 4C).

After 6 months later checking the panoramic radiograph displayed all the permanent tooth and tooth germs except #45, which are different stages of tooth development (Fig. 4D). The alveolar bone had no visible resorption.

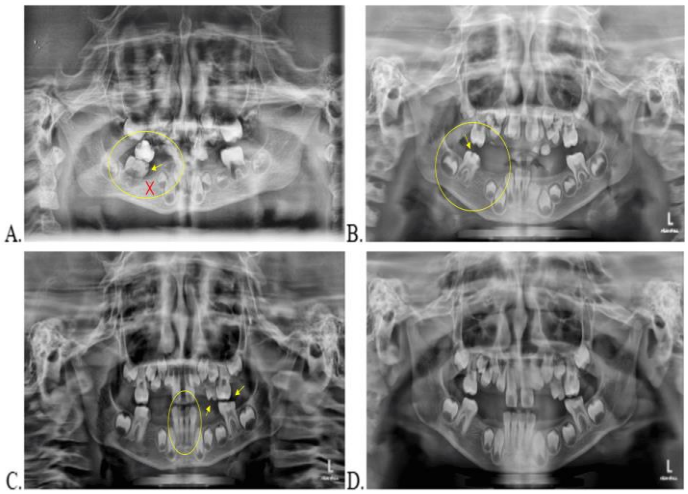

Fig. 4. Panoramic radiograph
(A-4.5 y.o-The yellow arrow indicates the radiopaque image on the mesial side of the crown of the mandibular right first permanent molar and yellow circle shows the local horizontal alveolar bone resorption. Red X sign says the absence of #45 tooth germs in the mandible bone [7]
B-5.9 y.o-The yellow arrow indicates the new radiopaque image in the coronal occlusal side of the mandibular right first permanent molar and the yellow circle says that alveolar bone was restored [7]
C-6.3 y.o-The yellow circle shows the eruption of the central and lateral incisors in the mandibular dental arch. The upper yellow arrow indicates that the left first premolar were erupted in the maxillary dental arch. The lower yellow arrow the new radiopaque image on the coronal occlusal side of the mandibular left first permanent molar
D-6.9 y.o-The alveolar bone had no visible resorption)

Diagnosis and treatment

We offered "Papillon-Lefevre syndrome" based on the patient's clinical sign, radiographic image, palm hyperkeratosis, edentulous, alveolar bone resorption and his pyogenic infection history. The treatment plan included oral hygiene modification, tongue ulcer treatment and a removable denture. During the oral hygiene modification some deciduous teeth fall prematurely by themselves. We eliminated the traumatic factor-the sharp enamel edge of an incisor that caused the tongue ulcer and the lesion fully healed within two months (Fig. 5a-10i) [7]. The patient goes to immune support treatment every two weeks and is supervised by the immunologist and goes to pediatrician and pediatric dentist every 3 months.

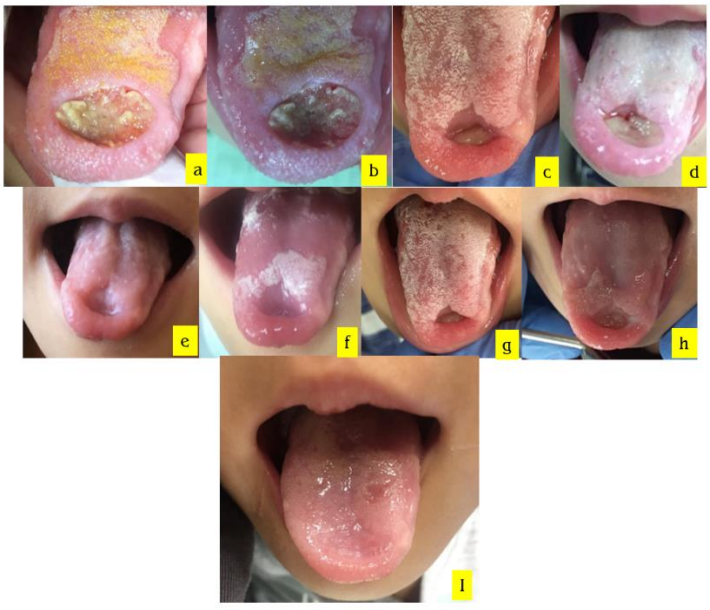

Fig. 5. The healing process of the tongue ulcer (a-first visit, b-7 days, c-14 days, d-21 days, e-28 days, f-35 days, g-42 days, h-49 days and i-70 days after treatment start, full recovered) [7]

By the age of 18, the patient will be considered for fixed prosthetic rehabilitation.

DISCUSSION

Papillon-Lefèvre syndrome is an infrequent genodermal syndrome characterized initially by two French physicians Papillon and Lefevre in 1924, in a brother and sister with palmoplantar hyperkeratosis accompanied with early onset periodontitis and premature loss of primary as well as permanent teeth. Gorlin et al. in 1964 added a third section of dural calcification to the diagnosis of this syndrome [1,8]. Papillon-Lefèvre syndrome has been categorized into three groups: diffuse, focal, and punctuated [4]. The hyperkeratosis of the plantar surface often spreads to the edges of the soles and in some cases onto the skin overlying the Achilles tendon and the external malleoli. We observed from patients the only palmoplantar hyperkeratosis, nail shape abnormal and transverse grooving [9, 10].The intraoral features such as the premature loss of permanent and deciduous teeth, mobility, and aggressive periodontal destruction were reported by Nickles et al. [11], Vassilopoulou and Laskaris [12]. The above mentioned clinical features were the same as our patient's clinical symptoms, which appeared as exposed tooth roots and the alveolar bone resorption on the panoramic radiograph. From 3 years following examination and radiographic results we were not observed alveolar bone resorption.Haneke advised to make a differential diagnosis of PLS using the following three criteria (palmoplantar hyperkeratosis, autosomal recessive inheritance, loss of primary and permanent teeth) from prepubertal periodontitis and Haim-Munk syndrome [4,13]. We confirmed the diagnosis, because our patient has the above mentioned 3 criteria.

According to published reports, the prevalence of PLS is approximately 1 to 4 cases per million people and with both parents as recessive carriers, there is a 25% chance of generating offspring with this syndrome [14,15]. The fathers of the patient had a full lip and palate cleft and he had chromosomal disorder. But the mother does not have any congenital symptoms. We occasionally observed some shortened tooth root in the panoramic radiographic of the patient's mother. But we can not confirm it was a symptom. In the dental history of the mother, her upper central incisors exfoliated when she was 8 years old. And her #11 implant denture was extracted 6 months ago due to peri-implantitis (Fig 2) and she was wearing temporary denture (ribbond teeth) until restored the alveolar bone of the premaxilla. PLS is not specifically recorded in our country, but the congenital developmental anomaly is registered among the newborns. So we considered that the PLS is referred to as ICD 0.80-0.89 (0.8%), or ICD 0.90-0.99 (4.2%) [6].

The pathogenesis of PLS is still controversial, but two hypotheses are more prominent nowadays. First, some patients suffering from PLS display a cellular immune fault with reduced chemotactic and phagocytic function of neutrophils and other granulocytes. Second, some pathogenic microorganisms like *Capnocytophaga gingivalis, Porphyromonas gingivalis, Actinobacillus actinomycetemcomitans, Peptostreptococcus micros, Fusobacterium nucleatum,* and spirochetes have been concerned as the causal agents for periodontal problems in PLS. We had no possible way to identify the pathogenic microorganisms in the periodontal pockets in this case. The patient is unable to produce immunoglobulin A, which aligns with one of the primary hypotheses regarding the pathogenesis of PLS [2,5,16].

Identification of PLS at an early age and a multidisciplinary approach can improve the prognosis of these patients. Skin lesions are commonly treated with emollients, salicylic acid, and urea. Oral retinoids including acitretin and isotretinoin have also been reported for the treatment of kertoderma [11,17]. Effective treatment of periodontal disease includes prompt institution of antibiotics with nonsurgical therapy, the modification of the patient's oral hygiene, extraction of primary teeth, and periodontal maintenance visits every 3 months. There is no consensus regarding the use of antibiotics; however, the prescription of antibiotics in combination with periodontal therapy has shown some benefit [17,18]. This case treatment consisted of the oral health modification and antibiotic therapy based on the results of the antibiotic susceptibility test of tongue ulcer. The patient is regularly enrolled in the recall system under pediatric dental supervision.

CONCLUSION

Papillon-Lefèvre syndrome can badly affect the psychological, social, and esthetic well-being of the patient at an early age, as it is a devastating disease process associated with cutaneous involvement and partial or complete edentulism. The dentist is usually the first to diagnose this syndrome due to the involvement of the periodontium.

DECLARATION

Conflict of Interest: There are no conflicts of interest.

Financial support: None

Competing Interests: The authors have no

competing interests to declare.

Ethical approval

We gained the written informed consent of the patient's parents to use his clinical information and radiological and photographic material for the publication. The Research Ethics Committee of Mongolian National University of Medical Sciences issued approval 24-25/06-02-08.

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