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Anesthesia Management of a Patient with Papillon-Lefevre Syndrome: A Case Report

Afshin Iranpour,[1,*] and Ata Mahmoodpoor[2]

[1]Department of Anesthesiology, Al Garhoud Private Hospital, Dubai, UAE
[2]Department of Anesthesiology and Critical Care Medicine, Faculty of Medicine, Tabriz University of Medical Sciences, Tabriz, Iran

[*]*Corresponding author*: Afshin Iranpour, Department of Anesthesiology, Al Garhoud Private Hospital, Dubai, UAE. Tel: +971-502567067, Fax: +971-44545197, E-mail: af6872@yahoo.com

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Abstract

Introduction: Papillon-Lefèvre syndrome (PLS) is a rare autosomal recessive trait; it often requires some interventions with general anesthesia because of the accompanied complications.
Case Presentation: We report a 19-year-old girl with palmoplantar hyperkeratosis who presented total loss of her teeth. She was candidate to mandibular bone graft and lower jaw dental implants under general anesthesia.
Conclusions: There are only a few studies about perioperative management of these patients; however, the anesthesiologists should consider a few important issues during pre-operative and intra-operative management.

Keywords: Papillon-Lefèvre Syndrome, Fiberoptic, Nasal Intubation, Dexmedetomidine, Dental Implants

1. Introduction

Papillon-Lefèvre syndrome (PLS), which is inherited as an autosomal recessive trait, is an extremely rare genodermatosis that first was described by two French physicians Papillon and Lefèvere. Palmoplantar hyperkeratosis and diffused rapid destruction of dental alveolar supporting bone are characteristics of this syndrome (1). It affects both the primary and secondary dentition before the age of 4. Exfoliation of teeth and rapid bone loss are the results of inflammatory response in the periodontium. The afflicted patients are usually edentulous which needs wearing complete dentures by their teen years because both sets of dentitions are affected (2). The exact immunologic abnormality of PLS is unknown. Microscopic changes include osteoclastic activity, marked chronic inflammation with predominant plasma-cell infiltration, lack of osteoblastic activity (3), and diminished neutrophil activity (4). Papillon-Lefevre Syndrome should be differentiated from Langerhans' cell histiocytosis (Histiocyto-sis X), Hypophosphatasia, and Haim & Munk Syndrome. Because of complications of this syndrome, some patients need to undergo interventions, which require general anesthesia. In this case, we report a female with Papillon-Lefevre syndrome referring to our hospital for mandibular bone graft and dental implants.

2. Case Presentation

A 19-year-old female was presented with palmoplantar hyperkeratosis from the age of 4 years and complete loss of teeth by the age of 14 which was the characteristic criteria for PLS. She was the second child born to apparently healthy consanguineous parents (cousins). Her older brother also suffered from PLS but two younger siblings were normal. Previous medical history showed that her permanent and deciduous teeth were lost after erupting normally.

On physical examination, there was diffuse palmoplantar keratoderma (Figure 1). Intraoral examination revealed complete edentulous ridges with normal overlying mucosa. In the panoramic view, severe maxillary and mandibular bone resorption along with bilateral pneumatization of maxillary sinuses were seen. Her chin was small but the other systemic examinations, routine laboratory examinations, chest X-ray, and skull X-ray were normal. She was subsequently provided with artificial dentures and she was a candidate for mandibular bone graft from her skull and dental implants of lower jaw. Complete blood count and liver function tests were also normal.

The patient was informed of the indication, and risks and benefits of fiberoptic nasal intubation under sedation. Preoperatively, after careful intravenous catheter insertion, she was premedicated with intravenous midazolam 1 mg and glycopyrolate 0.2 mg. Topical nasal vasoconstriction was achieved with oxymetazolin nasal drop and xylocaine spray 10% in both nostrils. Nasal oxygena-

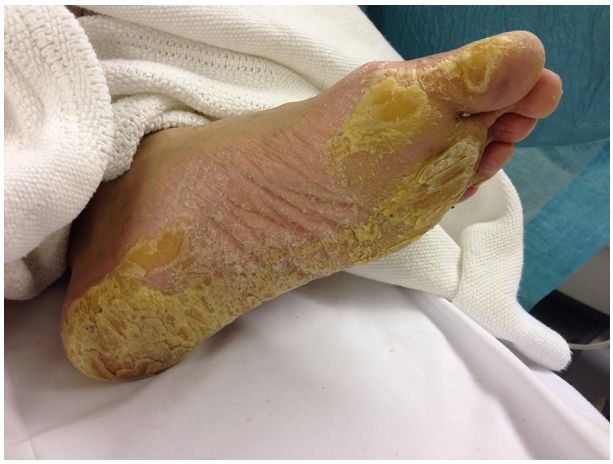

Figure 1. Diffuse Palmoplantar Keratoderma

tion through the nasopharyngeal airway with 100% oxygen was started 3 minutes before the nasal intubation. The patient received a loading dose of dexmedetomidine (Precedex) 1 μg/kg infused over 10 minutes, followed by maintenance dose of Precedex (0.4 μg/kg/h). Gently, she was intubated by fiberoptic. During the awake nasal intubation, the patient's SpO_2 remained between 97% and 100%. The heart rate varied from 82 to 96 beat/min. Fentanyl (1.5 μg/kg), propofol (2 mg/kg), and cisatracurium (0.15 mg/kg) were used for induction, and general anesthesia was maintained with Sevoflurane, 50% oxygen and 50% nitro oxide. Vital signs were stable during the surgery. Fentanyl (50 μg) and cisatracurium (2 mg) were repeated every 45 minutes.

At the end of the surgery, the patient was extubated when she was fully awake and shifted to post anesthesia care unit (PACU) and had a recovery without any complication. Informed consent was taken from the patient regarding the publication of information in the journal. The patient reviewed the case and gave written permission for the authors to publish the report.

3. Discussion

Papillon-Lefevre Syndrome is a rare autosomal recessive genetic disorder which is characterized by palmoplantar hyperkeratosis with precocious progressive periodontal disease since 1 - 4 years of age. The disease affects both males and females equally. Its incidence is almost 1 to 4 per million in the general population with a carrier rate of 2 to 4 per 1000 (5). Consanguinity of parents is demonstrated in about one third of cases. Palmoplantar hyperkeratosis sometimes extends onto the dorsal surfaces of hands and feet, resulting in a foul-smelling odor that usually manifests during the first 4 years of the life (6). Both the deciduous and permanent teeth are affected by severe inflammation and degeneration of periodontium, which starts at 3 or 4 years of age (3). Primary teeth frequently become loose and fall out by 4 years of age. After exfoliation, the inflammation decreases and gingival changes to healthy status. But, the process of gingivitis and periodontitis is usually repeated with eruption of permanent dentition, which results in exfoliation of permanent teeth without treatment. By the age of 14, most patients are usually edentulous (7). Dental implant-supported fixed prosthesis provides numerous advantages over the conventional denture in terms of cosmetics, functionality, longevity of the prosthesis, and patient satisfaction (8).

PLS is a very rare disease. There is little evidence concerning perioperative management of these patients. However, the anesthetic management has been classified into two parts: pre-operative and intraoperative management. As previously mentioned, this syndrome can lead to loosening of teeth and dental abnormality, and creates great preoperative challenges for anesthesiologist. In these cases, extraction of very loose teeth is recommended before anesthesia but we can use silk suture to help difficult airway management if there are many lost teeth. In addition, decreased lower facial height, retroclined mandibular incisors, upper lip retrusion, or maxillary retrognathia might be a reason for difficult airway management (9). As our patient had lost all permanent teeth, there was not the problem of difficult laryngoscopy; however, difficult bag mask ventilation might be resulted from complete loss of teeth and micrognathia.

On the other hand, palmoplantar hyperkeratosis can lead to difficulties in intravenous access; thus, this intervention must be performed with great precaution in patients with extensive skin lesions like our patient. In this case, we used nasal rout for intubation because of not interfering with the surgical field. After complete lubrication and using vasoconstrictor to avoid any tissue injury or bleeding in vulnerable tissues, we gently inserted fiberoptic bronchoscope. Liver function tests are recommended before any elective surgery in these patients because of accompanying pyogenic liver abscess (10). Moreover, diminished white blood cell function could result in an increased susceptibility to skin infection which should be considered during intravenous access; therefore, complete blood cells count is also recommended before elective surgeries. Nail changes are apparent in advanced cases and they are manifested by transverse grooving and fissuring which in severe forms might be interfered with correct pulse oximetry.

3.1. Conclusion

Bone graft and dental implant under general anesthesia are recommended for treatment of PLS patient. Therefore, anesthesiologists should be aware of the challenges associated with this syndrome and its comorbid conditions. Dental abnormality should be considered and evaluated preoperatively. These patients may have lost teeth at an unexpected age and hence, extra care should be taken to avoid dental damage during airway management

Footnote

Authors' Contribution: Afshin Iranpour: concept of manuscript, preparation of manuscript; Ata Mahmoodpoor: manuscript preparation, literature review.

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