Papillon-Lefèvre Syndrome: Prosthodontic Rehabilitation of Oral Function

Sir,

Papillon-Lefèvre Syndrome (PLS) is a rare inherited autosomal recessive disorder.1 It usually affects the skin and oral soft tissues, resulting in palmoplantar hyperkeratosis and early loss of teeth.2 PLS results in a severe degenerative periodontitis affecting both primary and permanent dentition. Deciduous teeth are lost as early as 4 years and most permanent teeth by the age of 14 years. The etiopathogenesis of PLS is not completely understood but it has been associated with mutations of CTSC gene located on 11q14.1-q14.3.3 We report a case with a complex disease presentation.

A 14-year girl reported with chief complaint of missing teeth, difficulty in eating, and esthetic concerns. A detailed history provided by the father, revealed an early loss of deciduous teeth by the age of 3 years. The patient had multiple extractions of permanent teeth due to periodontal breakdown. She was born with a patent ductus arteriosus (PDA) for which she was operated twice. The patient also suffered from cerebral palsy that resulted in bilateral talipes equinovalgus (TEV). The right foot was operated upon and bilateral ankle-foot orthoses (AFO) were provided. Parents had a non-consanguineous marriage and no other family member was affected. Extraoral examination revealed marked hyperkeratosis of the palms of hands and of feet with a generalised redness (Figures 1A and B). The nails also showed dystrophy and malformation. Upon intraoral examination, complete edentulism was observed with normal healthy mucosa and no inflammation. Older orthopantomogram (OPG) provided by the patient showed typical PLS finding of floating teeth (Figure 1C). For rehabilitation of oral function, conventional removable complete dentures with bilaterally balanced occlusion were selected. Patient has adapted well to the prosthesis and is on regular follow-up (Figure 1D).

No ultimate treatment exists for the prevention or management of periodontitis associated with PLS.1 Owing to young age and better motor skill, patients usually adapt easily and quickly to the prosthesis.4 The dentures, however, need to be replaced periodically to accommodate the growth of the patient. More recently, dental implants, with and without bone augmentation, have been used to provide implant-supported removable or fixed prosthesis as a definitive treatment modality. However, only a few cases have been reported in literature and long-term clinical outcomes need to be established.5

REFERENCES

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