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Familial Periodontitis Associated with Leukocyte Adhesion Deficiency Type I: A Case Report of Two Siblings

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ABSTRACT

Background: *Leukocyte Adhesion Deficiency Type I (LAD-I) is a rare autosomal recessive immunodeficiency characterized by defective neutrophil function due to mutations in the CD18 gene. It results in recurrent systemic infections and aggressive early-onset periodontitis.*

Case Presentation: *This report describes two novel cases of LAD-I in male siblings aged 10 and 15, both born to consanguineous parents. Both patients presented with severe periodontal inflammation, tooth mobility, cutaneous lesions, and a history of recurrent respiratory and gastrointestinal infections. Radiographic evaluation revealed generalized alveolar bone loss with a "floating teeth" appearance. Periodontal therapy included oral hygiene education, scaling and root planing, multiple extractions under general anesthesia, and antibiotic prophylaxis.*

Conclusion: *Early diagnosis and interdisciplinary management are critical in improving both periodontal and systemic health in patients with LAD-I. These cases underline the importance of dental practitioners being aware of oral manifestations of systemic immunodeficiencies.*

Keywords

Leukocyte adhesion deficiency, Periodontitis, Immunodeficiency, Pediatric dentistry, Case report.

Abbreviations

LAD: Leukocyte Adhesion Deficiency, CD18: Cluster of Differentiation 18.

Introduction

Systemic diseases have long been recognized as significant risk factors for the development and progression of periodontal diseases, particularly moderate-to-severe forms of periodontitis. Diagnosing these conditions requires a comprehensive, multidisciplinary approach, as highlighted by Armitage (1999) in his work on the classification of periodontal diseases [1]. He demonstrated that factors such as cardiovascular diseases, diabetes, and immune disorders, including leukocyte adhesion deficiencies, can interfere with the host's inflammatory response and adversely affect periodontal health.

During a workshop on systemic-associated periodontitis, it was emphasized that moderate-to-severe periodontitis is often one of the first clinical manifestations of immune deficiencies, such as those seen in Leukocyte Adhesion Deficiency Type I (LAD-I) [2]. This highlights the critical importance of early detection of clinical signs of periodontitis in managing systemic diseases, particularly in pediatric patients.

In the context of these systemic conditions, periodontal interventions must be tailored not only to manage the local inflammation but also to support the overall health of the patient. This holistic approach is especially crucial in the case of immune deficiencies, where early management of periodontitis can prevent severe complications related to the progression of infection.

Leukocyte Adhesion Deficiency Type I (LAD-I) is a rare primary immunodeficiency resulting from mutations in the CD18 gene, affecting the β2 integrin subunit essential for leukocyte adhesion and transmigration. The resulting impaired neutrophil function compromises the host's ability to combat infections and contributes to aggressive inflammatory responses [4,6].

Among its systemic manifestations, early-onset periodontitis is a prominent feature and often leads to premature tooth loss if left unmanaged. Oral signs may precede systemic diagnosis, making dental professionals vital contributors to early detection [3,7]. Herein, we report two novel familial cases of LAD-I presenting with severe periodontitis, and we review relevant literature regarding clinical management.

Case Report

Patient 1: A 10-year-old boy was referred for evaluation of gingival bleeding and tooth mobility. Born to consanguineous parents, his medical history included repeated hospitalizations due to lower respiratory tract and gastrointestinal infections. Extraoral examination showed ulcerative cutaneous lesions on the neck. Intraorally, generalized gingival inflammation, deep periodontal pockets (5–10 mm), and mobility of multiple teeth were noted. Orthopantomogram revealed generalized alveolar bone loss with a "floating teeth" appearance.

Patient 2: His 15-year-old brother presented with similar clinical signs, including severe gingival inflammation, tooth mobility, and extraoral necrotic skin lesions. He also had a history of recurrent systemic infections and multiple hospitalizations since early childhood. Radiographic findings mirrored those of his younger sibling.

Immunological evaluation had previously confirmed LAD-I in both patients. Periodontal treatment consisted of intensive oral hygiene instruction, full-mouth scaling and root planing, multiple tooth extractions under general anesthesia, and systemic antibiotic prophylaxis. Post-operative review after one month revealed notable improvement in gingival inflammation and reduced bleeding on probing.

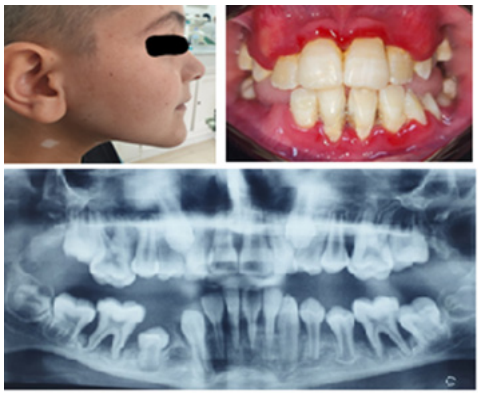

Figure 1: Extraoral and intraoral features of Patient 1 showing cutaneous lesions and generalized gingival inflammation. Radiograph shows advanced alveolar bone loss.

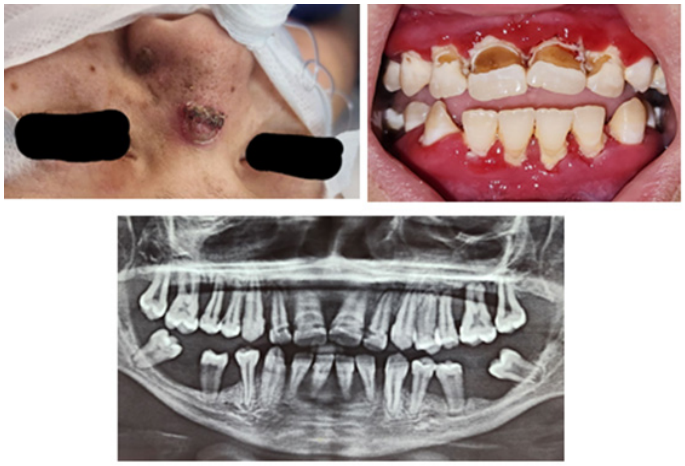

Figure 2: Clinical and radiographic presentation of Patient 2 with similar oral and cutaneous involvement.

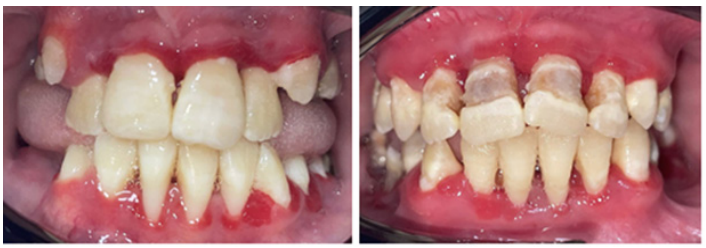

Figure 3: One-month follow-up showing improved periodontal condition post-treatment.

Results

Following the initial clinical and radiographic assessment, both patients underwent periodontal therapy as outlined. At the one-month follow-up, both siblings exhibited a significant reduction in gingival inflammation and bleeding on probing. The younger patient showed improvement in oral hygiene compliance and resolution of spontaneous bleeding. The elder sibling, while presenting with more advanced bone loss, also demonstrated clinical stabilization post-extractions and improved healing at the surgical sites.

Neither patient reported new systemic infections during this period, and no postoperative complications were observed. These outcomes suggest that despite their immunodeficient status, patients with LAD-I can respond favorably to carefully managed periodontal interventions supported by systemic care.

Discussion

The clinical manifestations observed in these siblings are consistent with previous reports on LAD-I, where impaired neutrophil adhesion leads to an inability to control oral microbial load and initiate effective wound healing [4-6]. The oral phenotype, characterized by aggressive periodontitis and early tooth loss, is frequently the earliest and most visible symptom [7].

Our cases contribute novel documentation of LAD-I-associated

periodontitis in a consanguineous Arab family. Early diagnosis by a dental practitioner is crucial to prevent further oral destruction and to guide referral for systemic evaluation. As emphasized by Armitage (1999), the association between systemic diseases and periodontal conditions underscores the importance of comprehensive patient evaluation [1].

Management requires a multidisciplinary approach. Although hematopoietic stem cell transplantation is curative [8], supportive periodontal care remains the standard in most resource-limited contexts. Adjunctive strategies including immunomodulation and local antimicrobial delivery have shown promise [2-11].

Conclusion

The presentation of these two related cases sheds light on the often-overlooked relationship between systemic immune deficiencies and early-onset periodontitis in pediatric populations. Severe gingival inflammation and rapid periodontal breakdown, particularly in the absence of poor oral hygiene or other typical risk factors, should prompt dental practitioners to investigate underlying systemic causes.

These siblings, both affected by LAD-I, exemplify how oral signs may precede a confirmed medical diagnosis and become the pivotal reason for referring the patient for immunological evaluation. Recognizing such patterns early is crucial, not only for the preservation of the dentition but also for the broader implications regarding systemic health, recurrent infections, and quality of life.

Management of periodontitis in immunocompromised patients requires more than conventional therapy. It calls for an adaptable and collaborative strategy that includes thorough infection control, supportive systemic care, and coordination with medical specialists. Long-term success in such patients depends on sustained follow-up, clear communication between caregivers and providers, and the empowerment of families through education.

Ultimately, this report reinforces the central role of the dental team in identifying systemic disease manifestations and contributing meaningfully to interdisciplinary patient care.

Consent

Written informed consent was obtained from the patients' guardians for publication of this case report and accompanying images.

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