ABSTRACT

Background Ligneous periodontitis or gingivitis is a rare periodontal disorder, secondary to plasminogen deficiency, characterised by nodular gingival enlargements and progressive destructive membranous periodontal disease.

Case Report We describe the early and successful dental management of a case of ligneous gingivitis secondary to plasminogen deficiency in a 6-year-old girl. The patient was referred because of a nodular asymptomatic gingival hypertrophy with ulceration around the eruption site of tooth 36, without any detectable tooth mobility. After non-surgical management of the lesion and strict follow-up, the first molar erupted completely, with no signs of bone and periodontal disease.

Conclusion Ulcerated periodontal lesions could represent the first signs of plasminogen deficiency. The early treatment is essential in preventing infections and the onset of a destructive periodontitis. The paediatric dentist may play a key role in early diagnosis and treatment.

Keywords Gingival hypertrophy; Ligneous periodontitis; Plasminogen deficiency; Ulceration.

Introduction

Plasminogen deficiency (PD) (OMIM 217090) is a rare autosomal recessive disorder characterised by impaired fibrinolysis during wound healing and subsequent accumulation of pseudomembranes on the affected sites [Schuster et al., 2007]. The most common clinical manifestation of the disorder is ligneous conjunctivitis, a form of chronic membranous conjunctivitis associated with fibrin deposits and “ligneous” (because of its wooden-like consistency) pseudomembranes on the eyelid surface. This may occur by itself or with similar lesions in other mucosal surfaces such as the nasopharynx, middle ear, larynx, vocal cords, tracheobronchial tree, and the female genital tract, indicating a systemic disease. The central nervous system can also be involved with development of cysts and/or hydrocephalus [Tefs et al., 2006]. Ligneous periodontitis or gingivitis is a rare periodontal disorder, secondary to PD, characterised by nodular gingival enlargements and progressively destructive membranous periodontal disease [Günhan et al., 1999] that can lead to rapid tooth loss despite of several treatment efforts [Baykul and Bozkurt, 2004].

This report describes the clinical manifestation and management of the early stages of ligneous periodontitis in a young Caucasian girl affected by severe PD.

Case report

A 6-year-old Italian girl was referred for paediatric dentist consultation because of the presence of pseudomembranous periodontal lesions. Since the age of two, she presented recurrent conjunctivitis and the development of membranous eyelid lesions, erroneously treated as chalazion. At the age of four, ligneous conjunctivitis was suspected and confirmed by plasminogen activity that resulted less than 29%. Genetic analyses were performed and a single mutation was found (K19E) on the PLG gene inherited by her father. She was treated with topical plasminogen eye drops. At intraoral examination, the patient presented a complete deciduous dentition, with the four first molars on their way to eruption. A nodular asymptomatic gingival hypertrophy with ulceration around the eruption site of tooth 36 was present (Fig. 1), without any detectable tooth mobility. The other permanent molars were also close to eruption without any signs of periodontal inflammation or ulceration. No radiographic signs of bone loss were found around the affected area, with the exception of a slight radiolucent enlargement around the crown of tooth 36 (Fig. 2). The patient, with PD and ligneous conjunctivitis, was diagnosed with ligneous gingivitis. The absence of destructive bone lesions did not indicate a need for invasive surgery. Scaling procedures were performed in order to avoid the
onset of a progressive destructive periodontal disease. Since then, a monthly follow-up was performed until all of her first molars were erupted completely, with no signs of bone and periodontal disease (Fig. 3).

She is now on a 6-month follow-up schedule, and is still under therapy with plasminogen eye drops.

Discussion and conclusion

This case report describes the clinical characteristics and management of the oral lesions of a patient affected by PD. She presents K19E PLG gene mutation, which is reported, in different studies, to be the most frequent genetic defect linked to PD [Schuster et al., 2007]. Although early manifestations are common in childhood, the disease first signs and symptoms are sometimes detected in the adult age. In this patient, ocular signs were the first to be found at the age of two, and no involvement of other mucosal sites was seen until the detection of pseudomembranous oral lesions at the age of six.

Many different oral signs, linked to PD, have been described as “ligneous periodontitis or gingivitis.” These heterogeneous signs range from gingival swelling, hyperplasia and mucous ulceration to severe periodontal destructive disease ending in partial or total edentulism [Sivolella et al., 2012]. To our knowledge this is the first case reporting pseudomembranous lesions secondary to PD around the eruption site of permanent teeth. A previous report referred to a case of ulcerated gingival swelling around deciduous molars in a 4-year-old female child, but no data were reported about oral lesion characteristics during the following mixed dentition [Gokbudget et al., 1997]. Wide heterogeneity is also reported about lesion localisation: some appear to be unilateral and many are bilateral with generalised gingival swelling and ulceration [Sivolella et al., 2012]. Micro-trauma has been related to the development of such lesions [Scully et al., 2001], and we believe that the possible aetiology of the single tooth involvement in this reported case is most likely due to an unreported micro-trauma and/or impaired tooth brushing.

Many treatment options have been reported in literature for these oral lesions, most of which with unsuccessful results [Baykul and Bozkurt, 2004]. In this case, a non-invasive treatment was performed with the aim of avoiding any periodontal infection of the membranous lesions and the onset of destructive periodontal disease. Due to the strict follow-up, no signs of periodontal disease or bone loss were reported, and the first molars erupted with no alteration in gingival and periodontal health (Fig. 3).

Ligneous periodontitis or gingivitis may be the first warning sign of a systemic pathology because of its easy detection. Young patients with pseudomembranous oral lesions should be evaluated for possible involvement of other mucosal sites, leading to the diagnosis of systemic diseases. Membranous periodontal lesions, in particular in proximity of tooth erupting sites, could represent the first signs of PD and pediatric dentists may play a key role in early diagnosis and treatment. The prevention of infections on pseudomembranous periodontal lesions is essential in avoiding the onset of a destructive periodontitis and severe bone loss around affected teeth.

References


