

Case Report

Ligneous Periodontitis in a Child with Plasminogen Deficiency

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ABSTRACT

Ligneous periodontitis (LP), a rare periodontal disease, is seen secondary to plasminogen deficiency and fibrin deposition. It is characterized by nodular gingival enlargements and progressive destructive membranous periodontal disease. It generally ends with the early loss of teeth. Defective fibrinolysis and abnormal wound healing is the main pathogenesis of this rare disease and should be considered different from other mucosal systemic disorders. In this case report, we describe the management of ligneous periodontitis and ligneous conjunctivitis developing secondary to plasminogen deficiency in a 6-year-old girl.

KEYWORDS: *Ligneous periodontitis, ligneous conjunctivitis, plasminogen deficiency*

INTRODUCTION

Ligneous periodontitis (LP) is characterized by the growth of gingival enlargement and periodontal tissue destruction that has not been fully defined. Amyloid-like material accumulation, plasminogen deficiency and deposition of fibrin is caused by this rare periodontal disease.^[1,2] LP generally starts in childhood, and conjunctival as well as gingival mucosa are its most affected sides.^[3,4] The etiology of LP is still not fully known. Possible causes such as autoimmune or hypersensitivity reactions, genetic disorders, trauma, viral or bacterial infections have been reported.^[4,5] In this case report, clinical and histopathological outcomes of a child who had LP and ligneous conjunctivitis (LC) is presented.

CASE REPORT

A 6-year-old female patient was referred to our department complaining of severe pain and gingival bleeding at the left maxillary posterior region in February 2015. She had presented with painless, solid, nodular fragile gingival enlargements involving right side of maxilla and bilateral mandible which had been presented about 2 years ago and they had been removed in another medical center. At intraoral examination, decayed first maxillary molar and nodular symptomatic

gingival hypertrophy, with ulceration around the eruption site of maxillary second molar, without any detectable tooth mobility was found [Figure 1]. The patient's growth and development was within normal limits, and similar problems were not seen in her family. Previous treatment which included gingivectomy and antibiotics were used for the treatment of oral lesions in another medical center. Unfortunately, these treatments were applied, but did not succeed. Radiological examination, decayed first maxillary molar and unerupted second maxillary molar was seen [Figure 2]. Since the second week of life, recurrent conjunctivitis has been seen as white membrane surrounding in the lower and upper eyelids mucosa [Figure 3]. Plasminogen analysis was done, and plasminogen deficiency was diagnosed due to hypoplasminogenemia which is named Type I plasminogen deficiency. But, any abnormalities were seen at haematological assessment, and biochemical tests such as liver function tests, serum proteins, azotemia and glycaemia were within normal limits. Hormonal values and kidney function tests were also within normal limits.

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Figure 1: In intraoral examination, ulcerated gingival swelling and membranous gingival enlargement is seen in left maxillary posterior region.



Figure 2: In radiological examination, decayed maxillary first molar tooth is seen and there are no problems in the bone.



Figure 3: Ligneous conjunctivitis is seen.

Under the effect of local anesthesia, maxillary first molar was extracted because of decay, the lesion was excised totally and biopsy was performed. Removed gingival tissues were examined histopathologically, and LP was diagnosed [Figure 4 and Figure 5]. After diagnosis of LP, the patient was referred to ophthalmology clinic because of recurrent conjunctivitis and LC was also diagnosed. In evaluation of patient, there was no cutaneous involvement or no relationship with any syndrome such as Ehlers-Danlos syndrome (EDS). Patient was still under regular control.

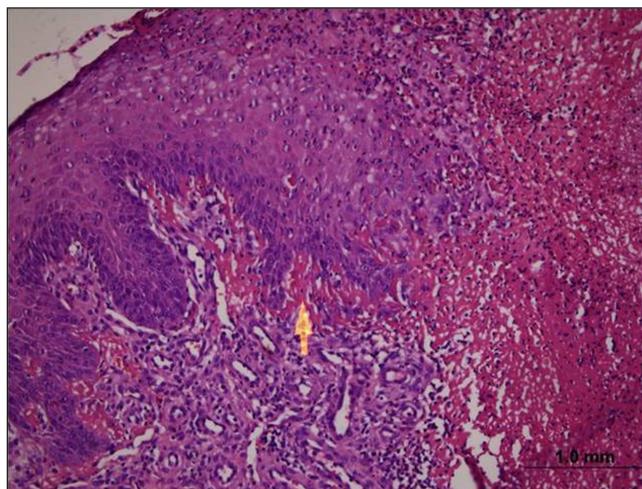


Figure 4: Early stage lesion shows hyperplastic changes in the epithelium, extensive intraepithelial neutrophilic infiltration and eosinophilic fibrinoid accumulation between the basal membranes (arrow). (HE, ×200)

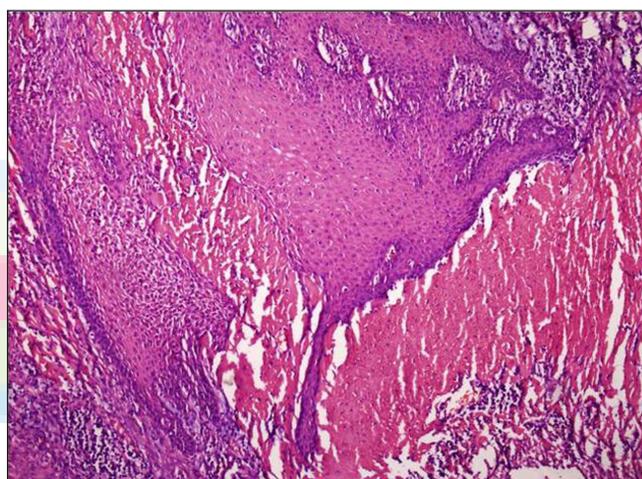


Figure 5: Late stage lesion reveals subepithelial amorph, eosinophilic, fibrinoid or amyloid like dense proteinaceous accumulation (HE, ×100).

DISCUSSION

Congenital plasminogen deficiency which is a rare autosomal recessive disease is clinically characterized by chronic mucosal pseudomembranous lesions depending on the sub-epithelial fibrin deposition and inflammation.^[6] LC is the most common clinical sign of this disorder which is an unusual form of conjunctivitis characterized by the development of firm pseudo membranes localized mainly on the tarsal conjunctivae.^[7] One of the extra-ocular symptoms of this disease is that it that may be associated either by itself or in similar lesions to the other mucosal surfaces such as middle ear, nasopharynx, vocal cords, larynx, tracheobronchial tree and the female genital tract indicating a systemic disease.^[8] In our case report, LC and LP which was seen in left maxillary posterior region is presented. Any pathological feature was not seen in other mucosal areas of the patient. Biochemical tests

such as serum proteins, liver function tests, creatinine and plasma glucose levels were within normal limits. Haematological examination showed no abnormalities, except plasminogen levels. Functional plasminogen analysis was done and revealed to be deficient.

The most characteristic features of this destructive periodontal disease are widespread membranous, nodular gingival enlargements in both the mandible and maxilla leading to rapid tooth loss despite several treatment attempts.^[1] In our patient's oral examination, widespread fragile nodular gingival enlargement, covered by a yellow-white membrane, partly covering the vestibular surfaces of the maxillary first and second molars was seen.

In such mucosal diseases, gingival biopsy is necessary for early diagnosis before the teeth are lost. Severe acute inflammation and irregular acanthosis of the epithelium with extensive fibrin leakage is the characteristic feature of this disease at the histopathological examination. This condensed fibrin accumulation lacks reticulin Fibers.^[1] Under local anesthesia, the lesion was excised totally and biopsy was performed. Removed gingival tissues were examined histopathologically; subepithelial amorph, eosinophilic, fibrinoid or amyloid like dense proteinous accumulation was seen, and ligneous periodontitis was diagnosed.

Despite a variety of surgical and periodontal treatment, attempts were unsuccessful in most of reported oral lesions.^[2,3,9] Therefore, treating such a disease was not promising until few years ago. Recently, Scully *et al.* suggested that gingival lesions can be controlled by topical heparin or intravenous purified plasminogen.^[9] Günhan *et al.* stated that systemic fibrinolytic and antithrombotic agents may prove more beneficial than local treatments, because the ligneous lesions tend to involve several mucosal areas.^[1] On the other hand, in some cases gingival lesions could be disappeared following tooth excision.^[10] In our case report, the gingival lesions became quiescent and disappeared after tooth extraction and lesion excision.

In conclusion, LP is a rare disease and seen as a result of plasminogen deficiency and subsequent fibrin accumulation. This disease occurs as pseudomembranous gingiva and can affect the eyes, or other organs. Dentists

should be familiar with this condition, since they might help in diagnosing this disease, with significant morbidity often missed by medical professionals.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil

Conflicts of interest

There are no conflicts of interest

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