# Clinical Radiographical and Dental Features of a Langerhans' Cell Histiocytosis Patient: Comparison of the Preliminary and Fifteenth Year

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## Abstract

Oral involvement in diffuse Langerhans Cell Histiocytosis (LCH) is a rare condition in children. Clinical symptoms are early loss of teeth, pain, gingival swelling and oral ulceration's. This case report aims to present and compare the preliminary and fifteenth-year clinical, radiographical and dental features a LCH patient and to suggest principles for long-term dental treatment planning of LCH. A 4.5 years old LCH patient (with Diabetes Insipidus), with the complaint of nutritional deficiency and chewing difficulties due to early, spontaneous exfoliation of the deciduous teeth was applied. Examinations showed that deciduous molars were mobile because of the destruction of supporting alveolar and basal bone. After dental treatments (extractions, restorative treatments etc.), removable space maintainers were applied. The patient could not attend the following appointments at all. After 15 years, bilaterally, molar open-bite has been detected and orthodontic treatment with orthognathic surgery has been indicated

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## Introduction

In 1953 Lichtenstein, was the first to describe Histiocytosis X term as a general name of a variable complex disease consisting of three clinical syndromes Eosinophilic Granuloma, Hand-Schuller-Christian and Letterer-Siwe[1]. In 1973, Nezelof et. al[2] proposed Langerhans' Cell Histiocytosis (LCH) term as an alternative to the Histiocytosis X since langerhans like cells were identified in the disease process. Therefore, the term LCH was approved to be used instead of Histiocytosis X term by the International Histiocyte Society, in 1987 [3].

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LCH is a rare disease and can be seen from early in life to elderly people with a peak incidence between 1-3 years of age[4].

Clinical presentations are variable and the restricted form of LCH may present erythematous skin-scalp rashes, organ involvement, cutaneous and lytic lesions, head and neck manifestations, lymphadenopathy and mild fever[5]. Bony lesions are usually single and are found in most patients. The skull is the most commonly affected structure and followed by long bones, flat bones, vertebrae and extremities[5].

The most common oral symptoms are the ulcerated mucosa lesions, periodontal lesions, gingival inflammations and bleedings, mobility and premature loss of teeth etc. in these patients. The mouth may be the primary site for the early manifestations. Symptoms that would cause a patient to get a consultation from a dentist vary from continuous oral lesions with pain, swelling, ulceration, gingival necrosis, dental abscess and painful swelling of the jaw [6]. In some cases, dentists are the

first to diagnose the disease[6,7], since loosening and premature exfoliation of deciduous teeth due to the destruction of supporting alveolar bone are the common signs of the disease[8].

This case report aims to present and compare the preliminary and fifteenth year clinical, radiographical and dental features of a LCH patient and to suggest principles for a long-term dental treatment plan of LCH.

**Case Report**

A 4.5-year-old, Caucasian boy applied to our clinic (Süleyman Demirel University Faculty of Dentistry, Department of Pedodontics) complaining of nourishing and chewing difficulties due to early, spontaneous exfoliation of the deciduous teeth in June of 2004. The past medical history of the patient revealed the presence of LCH and admission to the hospital two years ago, because of the scalp lesions and localized skin lesions around the perianal area. Diabetes Insipidus (besides LCH) was diagnosed in this admission. There was no previous history of surgery, chemotherapy, radiotherapy, or any other treatment. The only reason for treatment was for Diabetes Insipidus (DI).

Clinical examinations showed that the molar teeth 54, 55, and 75 were missing spontaneously. All the other deciduous molars were mobile, due to exposed roots and extensive profound dentin-cementum caries and the teeth 53, 62, 63, 73 and 83 had dentin caries (Fig.1a and 1b).

A radiographical examination showed multiple malpositioned deciduous and permanent teeth with loss of alveolar bone. The germs of teeth 14, 15, 24, 25, 34, 35 and 45 were absent. The alveolar portions of the jaws were destroyed and deciduous molars appeared like “floating in the air” (Fig. 1c).

Following the diagnosis, a consultation is made for the extractions of the involved deciduous molars, restorations of the teeth, and prosthetic rehabilitation and it was determined that the patient was in remission period.

Restorations of teeth 53, 62, 63, 73, 83 and extractions of the involved molar teeth 64, 65, 74 and 85 were performed under local anesthesia in separate, short visits (Fig. 1d). The teeth 84 had fallen spontaneously while the patient was chewing. During the extraction of the involved teeth, significant hemorrhage occurred and the extraction area was curetted carefully. Cephalometric analyses of the child were also done (Fig. 1f).

After the periodontal evaluation, prosthetic rehabilitation with partial, removable prosthesis was carried out (Fig.1e).

After 12 months of routine follow-up visits, the patient could not attend the following appointments at all because of moving to another city with the entire family, and subsequently, his father’s unexpected death.

After 15 years, coincidentally, contact has been established with the patient through social media. Afterward, the patient’s clinical radiographical and dental examinations have been performed in Akdeniz University Faculty of Dentistry, Department of Pedodontics. It was learned that the patient disuse his partial, removable prosthesis due to clinical complaints after one year it was made. Furthermore, in clinical and radiogra-phical examinations, it was detected that teeth numbers 14, 15, 24, 25, 26, 34, 35 and 45 were congenitally missing. Bilaterally, molar open-bite has been detected and an orthodontic/orthognathic treatment has been indicated. (Fig. 2 a,b,c). Cephalometric analyses (Dolphin Imaging 11.95 Premium, CA, USA) of the patient were also repeated after 15 years. (Fig. 2d)

Consequently, multidisciplinary dental management and long-term follow-up evaluation is required for these patients. Our patient has not been followed up for over 15 years. For this reason, bilaterally molar open-bite occurred, and the treatment is only possible with orthognathic surgery.

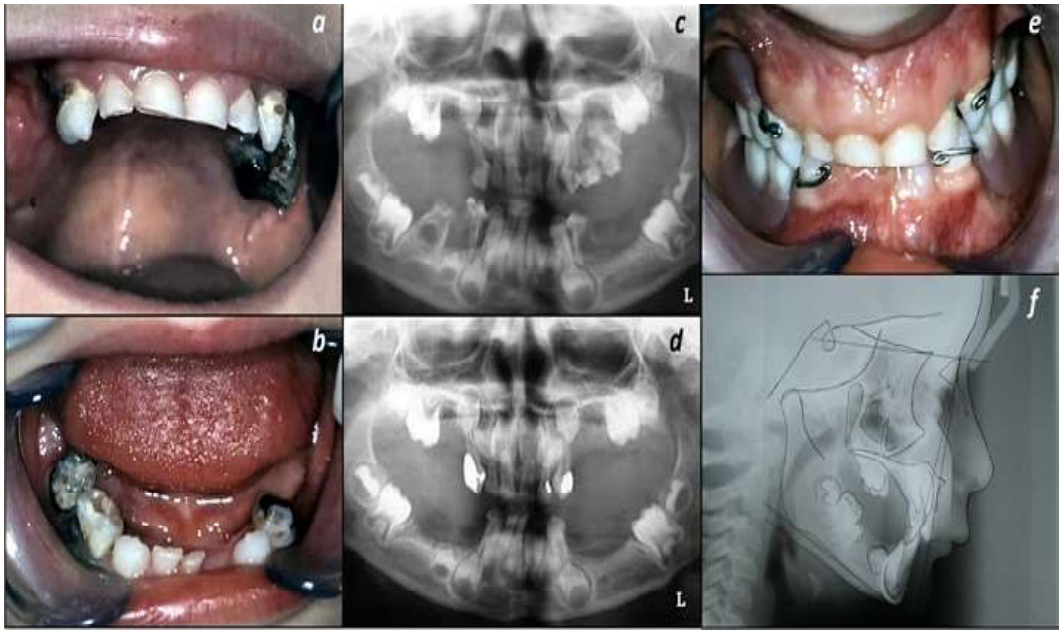


**Fig 1. a and b;** The clinical appearance of the child's upper and lower jaw. Both of the figures show deciduous molar teeth with extensive profound caries and exposed roots. **c;** On the orthopantomographic view of the patient deciduous molars appeared like "floating in the air". **d;** Radiographical appearance of the patient after dental treatments. **d;** Clinical appearance of the patient with partial removable space maintainers. **f;** In cephalometric analyses, SNA (82°), SNB (77,5°) and ANB (4,5°) angles were measured.

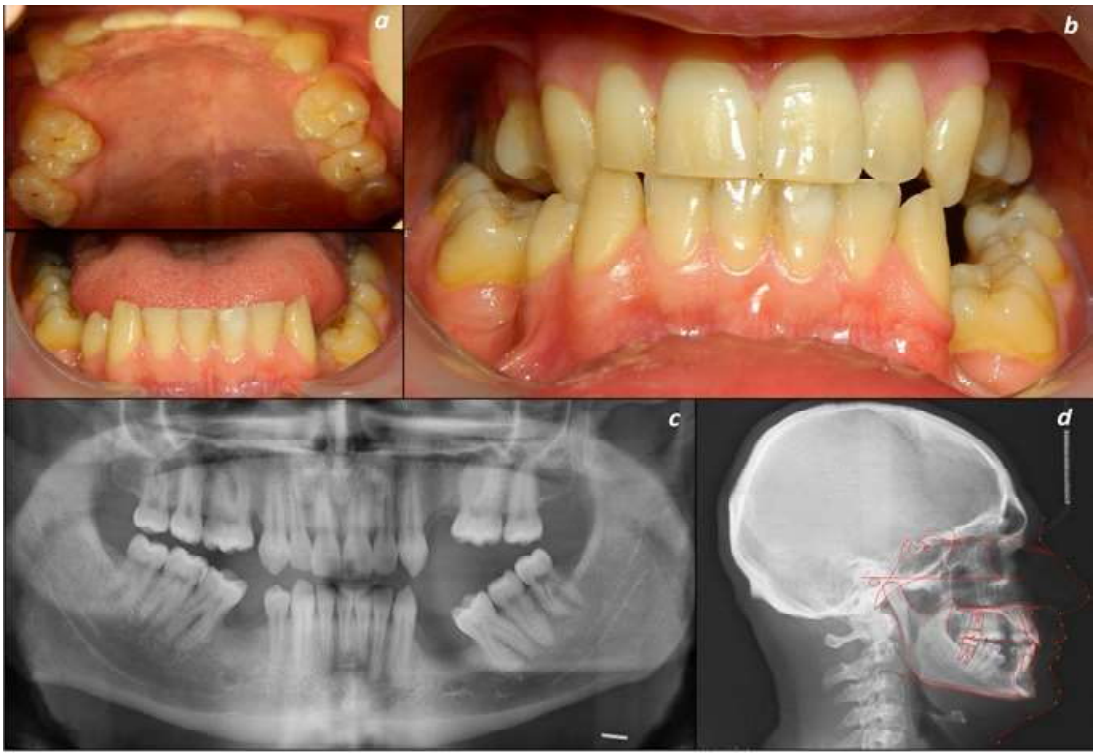


Fig. 2. **a;** clinical views of patients after 15 years. **b;** Bilaterally, molar open-bite was detected and an orthodontic treatment with orthodontic Surgery **c;** Orthopantomographic view of the patient after 15 years. **d;** Cephalometric analyses have been repeated and SNA (90°), SNB (87,3°) and ANB (2,8°) angles were measured.

## Discussion

LCH is a rare disease and its aetiology is obscure. However, it is generally considered to be an active disorder resulting from an immune activation[7]. LCH can affect multiple different organs. The skin is frequently the site of the presentation of LCH[5]. DI is the most common endocrinopathy[6]. In our patient, LCH started with the appearance of the erosive and ulcerative peri-anal, and skin-scalp lesions and followed by the development of DI.

Gingival inflammation and swelling are usually associated with cervical lymphadenopathy, which is the most common and the first manifestations of the disease[6]. Although head and neck involvements are observed frequently in young children, there was no lymphadenopathy in our case.

Accumulating studies and case reports mention that oral involvement is ranges between 10-77 % in LCH patients[5,6,8].

However, there was no oral symptom in our patient except for early loss of primary teeth. Premature eruption or an early loss of teeth arising from involvement of the mandible has been reported[8]. Radiographically, the appearance of posterior teeth “floating in the air” as a result of severe destruction of alveolar bone is a significant feature[6]. In our case, there were similar radiographic appearances in the posterior part of his jaw.

The differential diagnosis of the jaw lesions from a radiographic viewpoint included radiolucent defects as produced by some odontogenic cysts, tumors, granulomatous diseases, osteomyelitis, leukemia, malignant lymphomas, multiple myeloma, metastatic disease, periodontal and periapical lesions[6].

A diverse number of treatment approaches have been reported for LCH, according to extent of the disease[3,7,6]. These are surgical curettage, radiotherapy, chemotherapy and combined therapy. The patients who are in remission period as in our case and as, could be followed by routine controls without needing any treatment[9]. Many researchers have reported the risk of the reappearance of the event, which makes a long-term follow-up necessary[5,6].

LCH has effects on dental development and also long-term consequences should be observed[8]. Generally, extraction has been suggested for primary teeth with severe mobility or associated with lytic bone lesions[8]. In our case extractions of the involved primary teeth have been performed and partial, removable prostheses have been made in preliminary visits. Besides the functional improvement, positive psychological effects were observed in both patient and his parents.

## Conclusion

LCH often manifests itself initially in the oral cavity and dentists may play a very important role in establishing the early diagnosis of the disease. The frequent occurrence of oral manifestations necessitates the need for an interdisciplinary approach. Dental management of the syndrome with removable prostheses carries a big importance as well as medical treatment due to the functional, aesthetic and psychological aspects. Therefore, dentists and physicians must work together to confirm the diagnosis of the disease and to provide adequate treatment immediately for LCH patients.

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**Disclaimer:** None

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