Oral Manifestations of Langerhans Cell Histiocytosis in a 2½ Year Old Boy

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Introduction

Langerhans cell histiocytosis (LCH) is characterized by an abnormal proliferation of histiocytes of unknown cause. It is a rare disorder primarily affecting children. The incidence is estimated to be about 1:350.000 persons. LCH may affect various organs as solitary or multifocal granulomas including Letterer-Siwe syndrome, Hand-Schuller-Christian disease and Eosinophilic Granuloma. The latter is the most benign form of LCH. It shows a particular predilection for the head and neck region, and usually involves the skull bones. Oral lesions including gingival enlargement, oral ulceration, mobility of teeth and jaw pain are frequently the first and/or the only manifestations of the disease. General symptoms are irritability, loss of appetite and low grade fever.

Case

A 2 ½ year old boy was referred to the TAKO-centre by his paediatrician because of chewing problems and mobile molars. The diagnosis was not known at this moment. He had been treated for recurrent otitis with continuous secretion from right ear during the last year. Otherwise he was a healthy boy.
CT caput was planned.

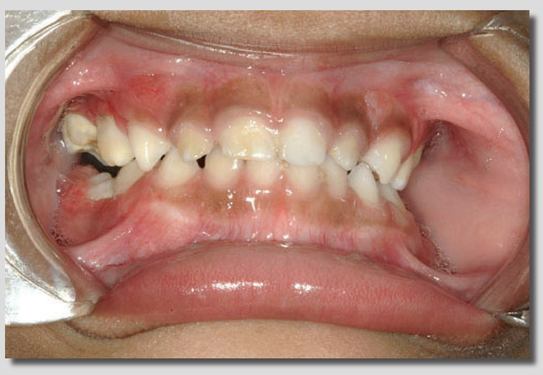
At the first examination photos and radiographs were taken. Intra oral examination showed all primary teeth present. Gingival swelling was seen in the upper and lower primary molar regions on buccal and palatal sides. The primary molars had pathological mobility. In the anterior region there was normal, healthy gingiva, no tooth mobility and no bone resorption.

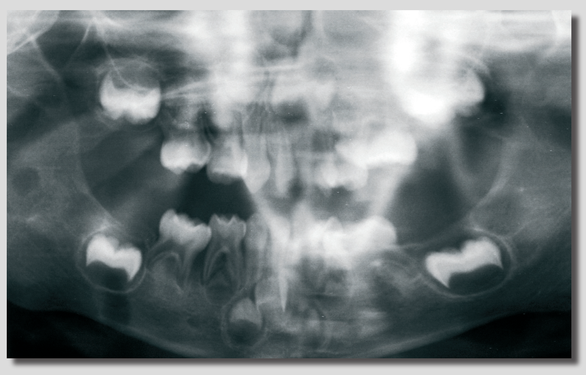
Radiographs revealed advanced bone destruction extending around the roots of the molars. Oral hygiene was very good, and they had used chlorhexidine mouth rinse on the tooth brush twice daily in the last two months.

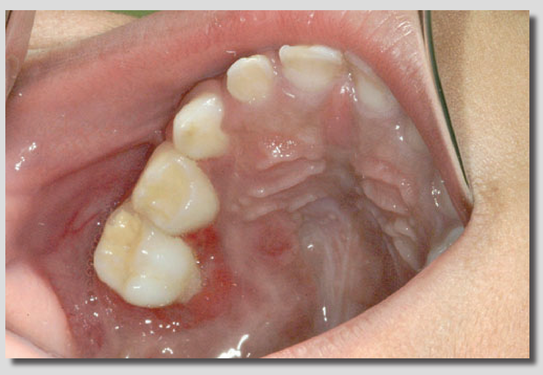
CT caput three weeks later showed bone destruction and granuloma in right temporal bone. Surgery was performed and histological investigation verified Langerhans cell histiocytosis, type Eosinophilic granuloma (EG). It was classified as a single system disease (only bone destruction), multifocal, because of skull and jaw affections.

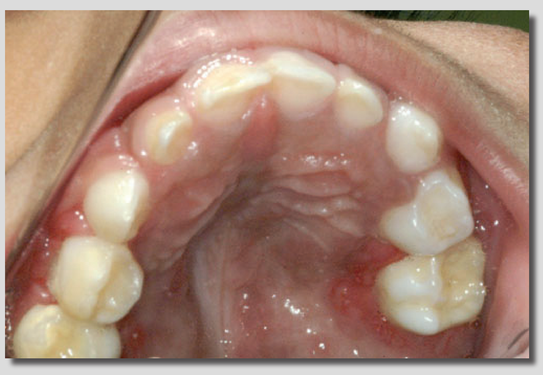
Treatment with steroids and cytostatic drugs started and followed a treatment plan over six months for patients with multifocal bone disease involvement.

First examination:









Oral treatment

The treatment plan after the first examination was to remove all second primary molars because of extreme mobility. However the medical advice was to postpone the extractions for some time, and observe if the medical treatment would lead to reconstruction of alveolar bone as expected in the skeleton.

So we did, and he was followed up by dental professionals with 2 weeks intervals in the beginning and 4 weeks intervals later on. Chlorhexidine dental gel was recommended for daily use at home.

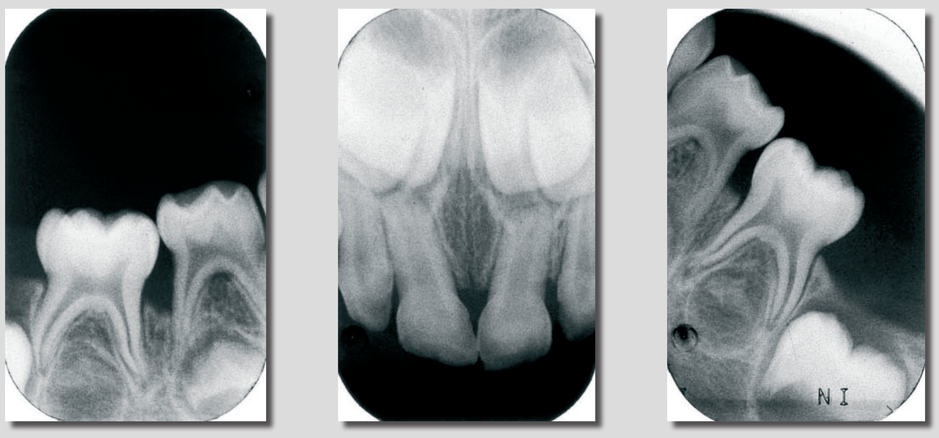
After six weeks we found the oral condition much better. He had no chewing problems and the molars were not so mobile.

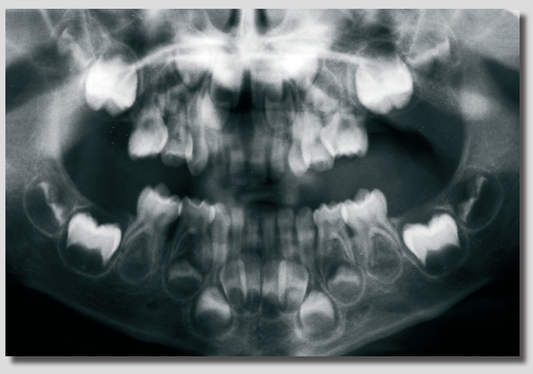
The boy responded well to steroid and cytotoxic therapy and the osteolytic lesion in temporal bone was healing. The general medical therapy was finished after 6 months and the oral condition was satisfactory. The molars had only physiological mobility and the gingiva was retracted but healthy.

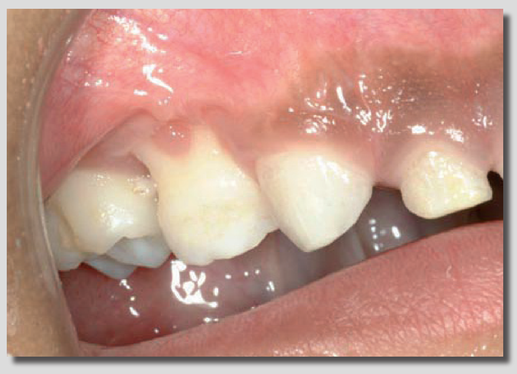
Treatment continued with follow up by dental professionals and use of chlorhexidin toothpaste twice daily.

Nine months after therapy onset, the result was very positive with some new formation of alveolar bone.

After nine months therapy:







Conclusions

According to the literature, the treatment in LCH with oral manifestations has been extraction of mobile teeth due to periodontal destruction. Experience in medical treatment has shown reconstruction of bone in skull and long bone with massive doses of steroids and cytostatics. In this case, we observed some new formation of bone around the roots in the molar region. So far all teeth are preserved and the little boy and his parents are satisfied. The condition requires optimal oral hygiene and long-term clinical and radiographic follow up.