Oral Manifestations of Histiocytosis X; A Review of 41 Cases

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Abstract

Langerhans cell histiocytosis (granulomatosis) is a rare disease of Langerhans' cells, and because of occasional involvement of the jaws it is important in the field of dentistry.

In this study, 41 cases of Langerhans' cell histiocytosis were collected from Pathology Departments of Mofid and Taleghani hospitals as well as Dental School of Shahed Beheshti University of Medical Sciences. 14 patients (34.1%) had oral manifestations, that is reported here.

Key words: Langerhans cell histiocytosis, granulomatosis, oral manifestations.

Introduction

Langerhans cells histiocytosis (LCH) is a rare disease of the dendritic cell system that was first introduced in 1953 by Lichtenstein. The disease can vary from a solitary bony lesion to a disseminated involvement of visceral organs, skin and bone.1,2 Although most cases are diagnosed during early infancy with a peak age between 1 and 3 years, adult patients are increasingly reported.3 Langerhans cell histiocytosis (idiopathic histiocytosis or Langerhans cell granuloma) is consisted of 3 types: eosinophilic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease. It seems that all the 3 types share a common process of multiplication of cells with a functional and structural view, similar to Langerhans cells. Since the identification of Langerhans cells as the multiplying cells, the term “Langerhans cell histiocytosis (granulomatosis)” first suggested by Risdall in 1983 and replaced the previous title.2,3

Occasionally the first manifestations of Langerhans cell histiocytosis occur in the mouth, in many cases being the only signs of the disease.2,4 The stimulus for proliferation is unknown, proposed theories have included metabolic, genetic, infectious, and neoplastic causes3,5 or a reactive process related to thymic dysplasia and deficiency of cellular immunity.2,5

Histologically, Langerhans cell histiocytosis is characterized by a proliferation of langerhans' cell with lobulated vesicular nuclei (coffee-bean appearance) associated with variable number of other inflammatory cells especially eosinophils.4

Patients and Methods

This descriptive study has done by observation and registration of data in the forms from the files available at Mofid and Taleghani Hospitals and also Dental School of Shahed Beheshti University of Medical Sciences from 1990 to 2000 and assess the variables of tooth mobility, pain, periodontal disease and bone involvement. Pain unrelated to the lesion was excluded.

Results

A total of 41 cases of Langerhans cell histiocytosis were detected. The patients' age ranged from 3 months to 40 years. Among the 41 cases, 14 (34.1%) were female and 27 (65.9%) were male. The mean age of female and male patients was 9.2 and 8.4 years respectively and the total mean age was 8.7 years.

14 cases (34.1%) including 10 (78.5%) males and 4 (21.4%) females had oral manifestation. The mean age for female and male patients was 13 and 14 with a total mean age of 13.5 years.

Among patients with oral manifestations, 13 (92.8%) had periodontal problems and tooth mobility. Tooth mobility as a distinct finding had been recorded in 6 cases (42.8%). There was pain in 3 cases (21.4%) and bony involvement in 10 cases (71.4%). 9 cases had multiple bony lesions (90%). All of these 10 cases showed involvement of the mandible associated with two cases of upper jaw involvement. Anesthesia of the lower lip was seen in one case (7.1%) and in two cases (14.2%) the disease had recurred in a different site.

Discussion

Sigala and Schepman have cited that the prevalence of oral involvement in the patients with histiocytosis X is 36%. Schepman also believes that dentists are the first clinicians to diagnose the disease in 16% of cases.2,3 In this study, a prevalence of 34.1% was recorded that is not considerably different from the previous reports.

Degenasia after evaluation of 29 cases of Langerhans cell histiocytosis of the jaw, reported

mandible as the involved site in 96.6% of cases. In the present study, involvement of the lower jaw was 71.45%, which is lower than the results of Degenasias's study but almost identical to the prevalence of 77% reported by Hartman.

Regarding periodontal disease, 79% of patients showed signs of periodontal disease in Grosky's study, whereas we found this to be 92.8%. As to the other variables, Ardekian showed that 92% of patients had pain and 7.3% had disease recurrence, whereas in our study it was 21.4% and 14.2% respectively.

Compared to the previous reports, low percentage of pain complaint was seen in the patients in our study.

References