Craniofacial Manifestations Of Eosinophilic Granuloma

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Citation

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Abstract

Purpose : The aim of the study was to present the cranifacial manifestations of eosinophilic granuloma with the review of the features , the choice of the method of treatment and the outcome of the disease.

Patients and Methods : We present 5 cases with the most common clinical presentations of intraoral swelling and loose teeth. The lesions were located at the maxilla and mandible . One patient had an ulcerative lesion at the temporal region of the scalp. The radiologic view showed lytic lesions at the scalp and maxilla-mandibular area. All of the patients had an incisional biopsy with the diagnosis of "Langerhans cell Histiocytosis".

Results : The choice of treatment was curettage in all patients and radiotherapy was combinated in one patient. Postoperative follow-up of the patients showed no recurrence

Conclusion : Intraoral lesions located at the maxilla and mandible associated with the loose teeth should be biopsied for the diagnosis of eosinophilic granuloma.

INTRODUCTION

Eosinophilic granuloma (EG) of bone may be defined as a benign , well-localized solitary or multiple osseous lesions characterized by large number of eosinophils and Langerhans type histiocytes([1],[2],[3]). It is first described by Lichtenstein and Jaffe in 1940 ([1],[4],[5]). Independently of them , Otoni and Ehrlich described cases showing the same type of the lesion under the name of “solitary granuloma of bone”. It is now known that the disease process may be present not only in one bone but in several or even many bones , so the designation “solitary granuloma” is already inappropriate ([5]).

Eosinophilic granuloma is classified as one of a triad of nonlipid reticuloendothelial disorders; Histiocytosis X disease which may be described as a nonneoplastic , proliferative disorder of histiocytes of unknown aetiology ([6],[7]). EG differs from Hand-Schüller-Christian disease and Letterer-Siwe disease in that it is a less agressive disease process, confined to bone, occurs later in life and demonstrates an abundance of eosinophils ([1],[2],[4],[5],[6],[7],[8]). Hand-Schüller-Christian disease which is the chronic disseminated histiocytes includes bone lesions, Diabetes Insipidus and exophtalmia ([1],[7]). Another dimension of the disease spectrum is an acute systemic illness named as Letterer-Siwe disease. It occurs mainly in infants and young children with a rapidly fatal course manifesting itself in multiple organs ([1],[7]).

EG is the least severe and most common form of Histiocytosis X. It can affect any bones with the exception of those of hands and feet ([2],[3]). It is more common in the pelvis, ribs, skull, vertebrae, facial bones, femur, tibia and jaws ([1],[2],[5]).

The jaws are affected in 10%-20% of all EG cases. The posterior mandibular region is the most frequently affected site ([9]).

MATERIALS AND METHODS

We would like to present 5 cases whose ages range from 4 years to 44 years.The most common clinic presentation was intraoral swelling and mass.Some of them also complained of loose teeth, tenderness and fetor oris.One patient had an ulcerative lesion located at the temporal region of the scalp.The laboratory findings were normal.The panoramic radiograph showed lytic lesion in the mandible.The craniography of the patient with the temporal lesion showed cranial bone destruction. Four year old patient had nuclear bone scanning with Tc 99m which revealed increased uptake of the radionuclide in the mandible.All of the patients had an

incisional biopsy with the diagnosis of “Langerhans cell Histiocytosis”. A visceral ultrasonography and radiological skeletal survey showed no evidence of multifocal soft tissue or skeletal involvement. The choice of treatment was curettage. Postoperative follow-up of them showed no recurrence.

Figure 1

Table 1: Profiles of 5 patients with eosinophilic granuloma

Patient	Age	Localization	Treatment
C.E	4	Right corpus of mandible	Curettage
A.Y.	44	Left temporal bone and left parasymphysis of mandible	Curettage and Radiotherapy
M.S.	28	Right maxilla	Curettage
R.S.	19	Bilateral corpus of the mandible	Curettage
L.H.	43	Right maxilla and left corpus of the mandible	Curettage

Figure 2

Case 1a : Preoperative panoramic radiograph of eosinophilic granuloma located at the right corpus and angulus mandible.

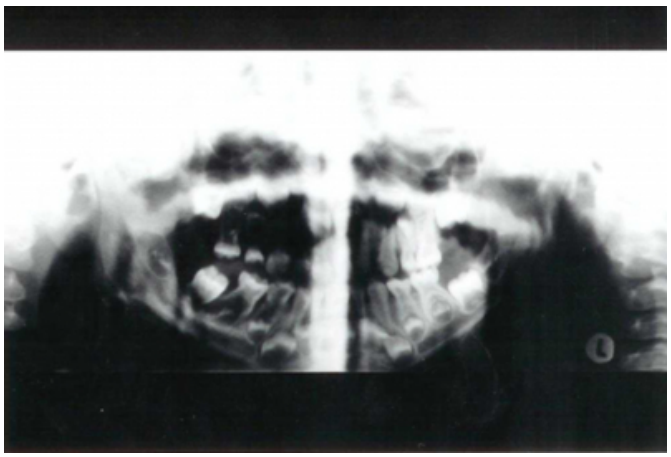


Figure 3

Case 1b : Panoramic radiograph of the mandible after curettage of the lesions.

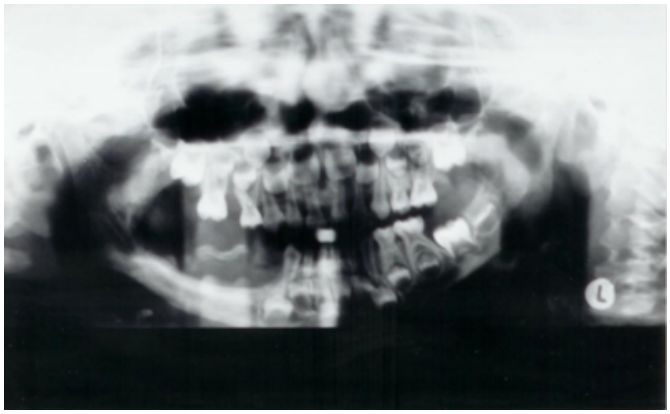


Figure 4

Case 2a : Ulserative lesion located on the left temporal area.

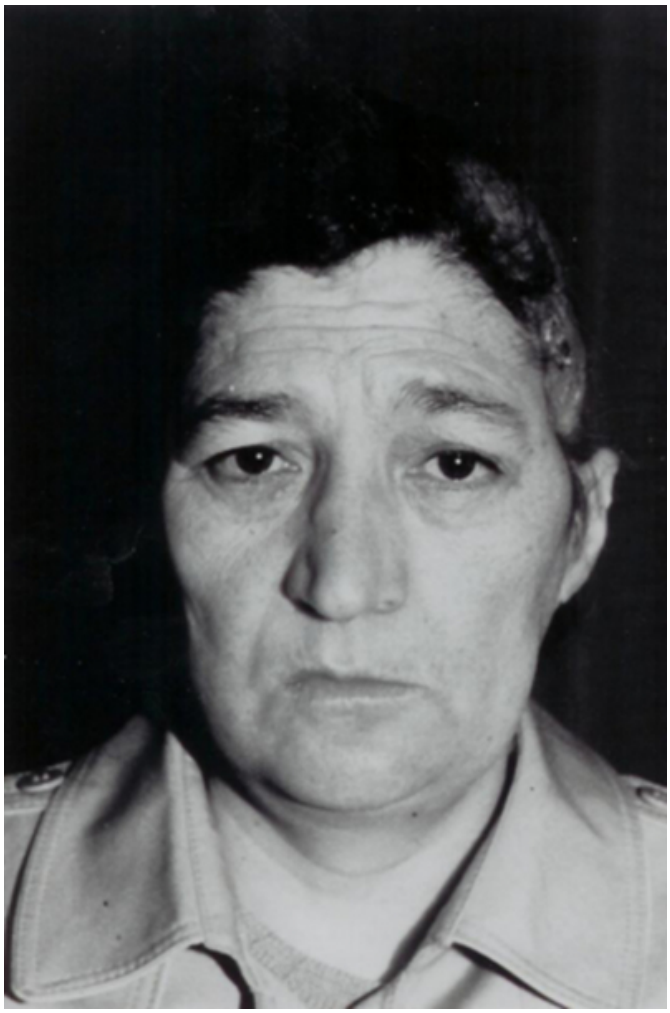


Figure 5

Case 2b : Ulserative mucosal lesion which caused hypermobility of the teeth located at the left parasymphysial mandibular region.

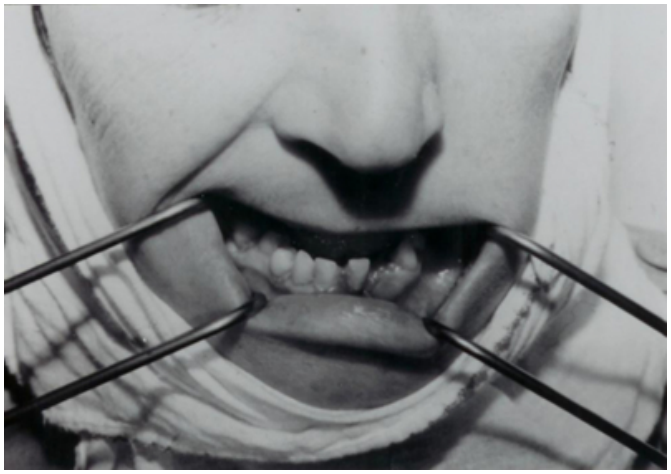


Figure 6

Case 3: Vegetative mucosal lesion located at the maxilla.

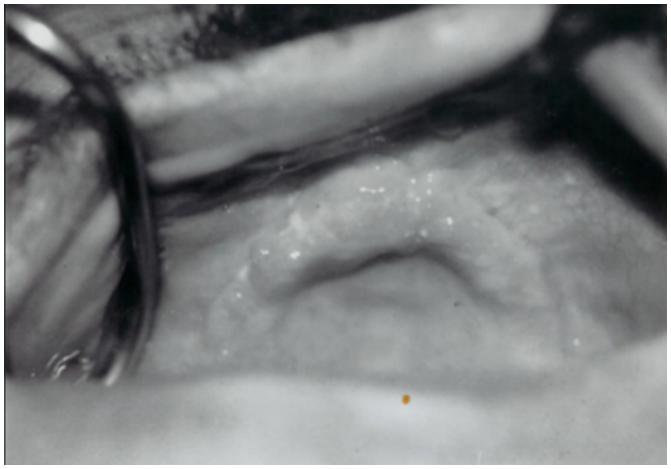


Figure 7

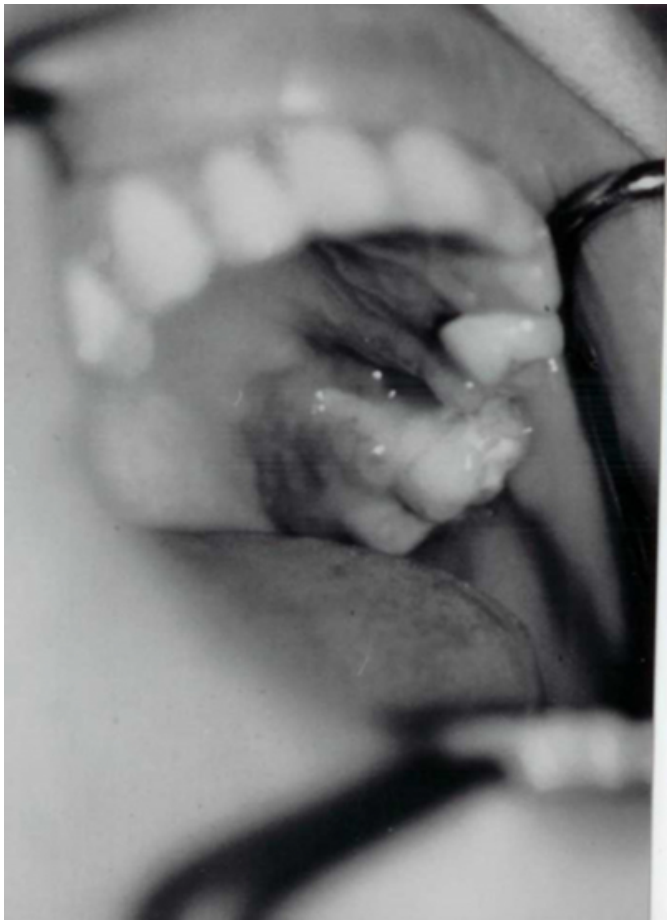
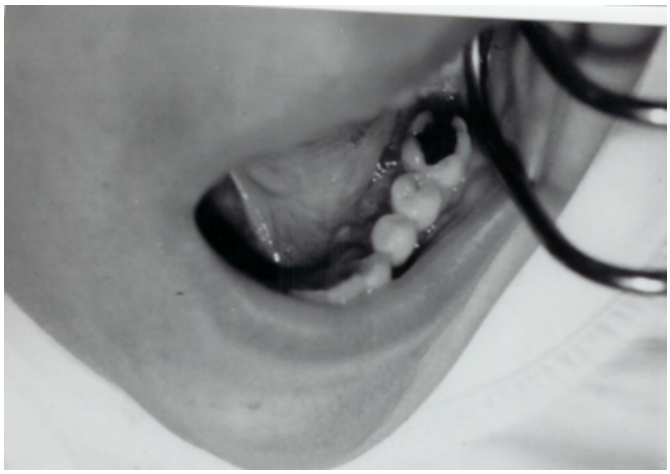


Figure 8

Case 4 & 5 : Mucosal lesion located at the left maxiller molar area and the left corpus of the mandible.



DISCUSSION

The etiology of Eosinophilic granuloma is obscure. It is neither hereditary nor a familial disorder ($_2$). It was once thought that the disease was a disorder of lipid metabolism because of the frequent finding of foam cells, but this is

generally not accepted now ([10]). It is thougt to be Langerhans cell origin which is an accessory immune cell that serves as antigen presenting cell . Other theories are that the cause of infection, allergic, trauma and neoplastic. However an infectious agent has not been identified and trauma serves only to call attention to a previously existing lesion ([8,9]).

Eosinophilic granuloma of bone may be characterized by localized pain, tenderness and swelling over the affected area and causing in some cases only a mild generalized reaction of fever, malaise, anorexia and headache ([1,3]). EG with oral involvement may present with gingivitis, loose teeth, oral ulceration and fetor oris.

The oral characteristics of this disease represent an entirely different clinical picture from other bones.This is due to the anatomy of the the structures involved, the presence of teeth and to the ever present of bacteria in the mouth which predispose to secondary infection ([3]). Many patients have loose teeth extracted and sockets fail to heal ([10]). It may stimulate advanced periodontoclasia due to marked resorption of the alveolar bone ([3]).

In lesions of the skull, a swelling over the affected area is often present which is moderate consistency and sometimes not painful at all ([3]).

Pulmonary involvement may be present in 20% of patients with EG of bone and at least 28% of these will have spontaneous pneumothorax ([10]).

In a differential diagnosis,one must consider multiple myeloma, metastatic tumor, inflammatory lesion,osteitis fibrosa cystica, solitary bone cyst, tuberculosis, syphilis, osteogenic sarcoma, giant cell tumor, Ewing's sarcoma and osteomyelitis ([3]). These conditions may be ruled out by taking a biopsy specimen,doing the necessary laboratory studies and by noting the variations in clinical symptoms and radiologic appearance of these lesions.

EG of bone is a lesion of the medullary cavity which roentgenographically shows erosion, expansion and at times perforation of the bony cortex to invade the surrounding soft tissues ([3]).The typical radiographic presentation of EG is a well-demarcated, radiolucent lesion , cortical bone erosion is frequently encountered and may lead pathologic fractures ([7]). In the jaws , root resorption is frequently seen.

The diagnosis can be made accurately only by obtaining a biopsy of the lesion.

Farber and Albers and their colleagues summarized the histopathologic changes ; While early lesions include large number of eosinophils, more nature lesions consist of lipoid-like histiocytes and late lesions show fibrous tissue changes with few eosinophils ([3,7,8]). Charcot-Leyden cyrstals were reported in 97% of cases ([6]).

Microscopically as the lesions age , the quantity of eosinophils decreases([7]).

The laboratory findings are neither consistent nor characteristic. Most studies are usually normal.There may be slight leucocytosis and eosinophilia , but this does not always occur ([3,7]). The accepted treatment for solitary EG includes curettage, low-dose radiaton, chemotherapy and steroid injection. All of these have been successfully used alone or in combination.

Accessible lesions are best treated by currettage which can be usually be done intraorally ([2,6,8]). In recent years radiotherapy has been limited to those patients who have extensive , rapidly progressing bone lesions and lesions not accessible to currettage or where operation will result in a gross disfigurement ([2,6]).

Prognostic factors are related to the number of organs or systems involved. EG of bone (polyostotik or monostotik) has the best prognosis out of the triad of Histiocytosis X ([1,6]).

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