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Eosinophilic Granuloma of the Mandible: Is a Conservative Treatment Sufficient for Local Disease Control?

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Abstract

Abstract: Eosinophilic granuloma is the most benign and common form of the Langerhans cell histiocytosis, a rare proliferative disease that can affect single or multiple organs. In the quite common head and neck manifestation these lesions can be confused in the beginning, with other bone diseases such as odontogenic cysts, periodontal disease, or malignancies. Treatment varies depending on the size, number, localization of the lesions, and patient's general conditions. The authors describe here a patient of a single lesion of eosinophilic granuloma localized in the posterior mandible region treated with a very conservative surgical approach in a patient with poor socio-economic conditions. The authors performed teeth extractions, an excisional biopsy and open curettage and after a follow-up of 24 months without partial mandible resection and reconstruction time the lesion had healed. The clinical situation confirmed that, in carefully selective patients, a conservative approach could be a useful therapeutic opportunity.

Langerhans cell histiocytosis (LCH) is a rare idiopathic disorder of the reticulo-endothelial system with abnormal proliferation of the bone marrow derived from Langerhans cells that usually reside in the skin and lymphnodes, along with leukocytes, such as eosinophils, lymphocytes neutrophils, and plasma cells.1 One of the possible manifestations is the eosinophilic granuloma. It is more commonly found in children, rare in the adult population, and additional multifocal involvement is even rare. Usually, it is localized in the bones as solitary or multifocal bone defects. Head and neck manifestations are quite common in LCH (69–73% of patients), involving the jaws in 7 to 10% of patients.2 Lesions might resemble periodontal diseases, odontogenic cysts, ameloblastoma, and malignancies; biopsy is mandatory for the diagnosis; information about the location, size, and extension is provided by computed tomography and positron emission tomography.3,4 The severity and prognosis depend on the type and extension of the organ involved. The treatment of Langerhans cell histiocytosis is various and the literature reveals a lack of consensus. We report below a patient of an eosinophilic granuloma of the mandible in a young patient treated in a conservative way with a biopsy followed by a vigorous curettage of the lesion.5

Keywords: Eosinophilic granuloma; histiocytosis; Langerhans cell; mandible

Clinical Report

A 28-year-old man in poor socio-economic conditions was referred to the Division of Maxillofacial Surgery, Città della Salute e della Scienza Hospital “Molinette,” University of Torino, Italy, for an evaluation of pain and swelling he was suffering of the right mandible. Clinical examination revealed very poor oral hygiene, a modest swelling of the alveolar process and the nearby gingival area, extended from the right mandibular premolar to the third molar, and teeth mobility from 4.4 to 4.7. No regional lymphadenopathy was observed. Sensibility alteration of the inferior alveolar nerve was reported. The panoramic radiograph showed an undefined osteolytic lesion at the right mandibular body (Fig. 1A). In the differential diagnosis of the lesion we considered it to be either a bony cyst, chronic osteomyelitis, ameloblastoma, sarcoma, or giant cell granuloma. An incisional biopsy was performed and a

diagnosis of eosinophilic granuloma was rendered. The histolopathological evaluation revealed the presence of eosinophilic and histiocyte cell, lymphocytes and plasma cells, and immunohistochemical positivity to CD1a and S100 (Fig. 1B). A computed tomography scan was performed and identified a significantly destructive bony lesion with interruption of both the cortical vestibular and lingual plate extending from the first incisive to the mandible angle involving the alveolar nerve canal with complete disappearance of the spongious bone (Fig. 1C). Consistent with the literature, a TC-PET total body with (18F) FDG was performed to better define the margins of disease and to evaluate the possible multiorgan interest. The examination revealed uptake of radiotracer in the right mandibular portion (SUVmax 9.05), in the anterior mandible portion (SUVmax 7.99) (Fig. 1D), and in the greater curvature of the stomach, without correlation to any type of clinical symptoms. We did not treat the lesion in a conservative way using corticosteroids and vinblastine because the patient refused a continuous therapy. The right treatment for this lesion could also be a partial mandibular resection, using an extraoral approach, with a large 2.4-mm reconstruction plate position under general anesthesia but the patient also refused the recovery. We took in consideration a conservative surgical treatment that the patient approved, consisting of an excisional biopsy of the lesion followed by vigorous curettage and drilling of the residual cavity, to be sure not to leave any part of the lesion, under local anesthesia, accompanied by extraction of the floating teeth (4.5 and 4.6). The postoperative period was uneventful. After 15 days a new panoramic radiograph was taken which showed the presence of an important bone defect due to the removal of the lesion and the drill of the cavity (Fig. 1E). No evidence of recurrence was observed at the 24-month follow-up. The new panoramic radiograph revealed modest signs of bone regeneration (Fig. 1F). Clinically, the epithelialization of the cavity was complete and without dehisce (Fig. 1G). A satisfactory, postoperative, functional result was obtained which correlated to the patient's condition and decisions.

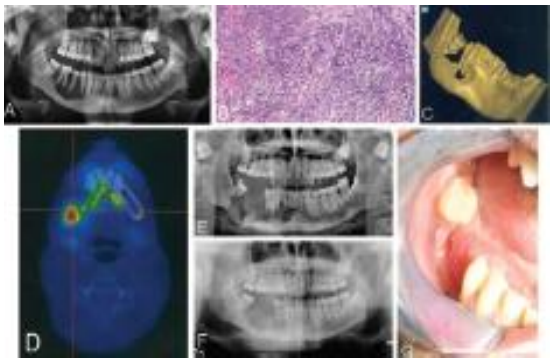

FIGURE 1. (A) Preoperative panoramic radiograph showing a not well-defined osteolytic lesion at the right mandibular body involving the apices of both the canine and the erupted third molar, with a typical floating tooth appearance. (B) Histological and immunohistochemical staining of specimens from the mass. H&E stain (magnification □20): presence of eosinophilic and histiocyte cell, lymphocytes and plasma cells. (C) Preoperative maxillofacial computed tomography scan with 3D reconstruction of the right mandible. The lesion extends to the right incisive involving the canal nerve. Destructive bony lesion with interruption of both the cortical vestibular and lingual plate extending from the first incisive to the mandible angle involving the alveolar nerve canal with completely disappearance of the spongious bone. (D) Positron emission tomography, red crosshair showing the lesion localized in the mandible. (E) Postoperative panoramic radiograph obtained at 15 days postoperatively demonstrating the residual cavity. (F) A panoramic radiograph at 2-year follow-up showing modest new bone formation. (G) Intraoral view showing epithelialization at 2-year follow-up.

Discussion

Eosinophilic granuloma is a severe manifestation of the Langerhans cell histiocytosis that can affect single or multiple bones, characterized by local proliferation or disseminate forms of dendritic cells of bone marrow. A recent study suggests that some patients may derive from circulating plasmacytoid monocyties rather than from epidermal Langerhans cells.

Langerhans cell histiocytosis is a rare disease, first described by Lichtenstein in 1953 with the term “Histiocytosis X” which comprises 3 different diseases with a common origin, stemming from a unique histiocyte with cytoplasmatic inclusions known as X bodies, with the X denoting the unknown etiology: Eosinophilic granuloma (LHC of bone, solitary or multiple), Hand–Schuller–Christian disease (bone lesions, exophthalmus, diabetes insipidus), and Letterer–Siwe disease (multiple organ LCH in early age). The term was changed to Langerhans cell histiocytosis by the Writing Group of the Histiocyte Society in 1985 to more accurately highlight the histologic characteristics and cellular origin of the disease.5 This rare disease, with an incidence of 8 to 9 patients per million/year, usually affects children and young adults, with men affected 1.6 to 1.7 times more often than women.6 As shown in the literature, head and neck involvement is common, ranging from 55 to 80% of patients, and the jaw is the most frequently affected site, in 10 to 20% of all EG patients, in particular the posterior mandible region.7 The disease may present in many different ways ranging from none to swelling, pain, tooth mobility, limitation of mouth opening, ulceration, gingival inflammation, and even pathological fracture. Facial asymmetry, malaise, and fever have also been reported.2 In the present patient, a 28-year-old man in poor socio-economic conditions reported mandible swelling and mobile teeth. When he was referred to our department we detected a modest swelling of the alveolar process and the nearby gingival area, extended from the right mandibular premolar to the third molar, and teeth mobility from 4.4 to 4.7. Routine diagnosis is usually made on the basis of histological examination and the presence of S-100 and CD1a by immunohistochemical analysis.1 The cells usually present abundant cytoplasm, indistinct borders, and oval to reniform shaped nuclei, often arranged in sheets and may be admixed with various numbers of eosinophilis.

Treatment of LHC has changed over the years, depending on whether the lesion was single or multiple and on its dimension, but there is no still consensus in the literature. In the literature medical treatments are suggested such as chemotherapeutic treatment, using intralesion injection of corticosteroids or vinblastine 3 and radiation therapy,8,9 but the patient did not want a continuous therapy. Since that the dimension of the lesion was consistent, we decided to approach this lesion in a surgical way. We proposed a partial marginal mandible resection using an extra-oral approach with a large reconstruction plate position to avoid pathologic fractures in the future. An autologous bone reconstruction would be the gold standard treatment to achieve good functional and aesthetic results, but the patient refused the recovery. Since in the literature some studies report the healing of these lesions after a single curetting biopsy and considering the patient's decisions and his poor social condition, the conservative surgical approach appeared the right choice in this patient.5 We had no recurrence at the 24-month follow-up. Maybe, a medical treatment could be sufficient for the control of a localized disease, but a conservative surgical approach can be a good alternative choice. A not completed excision of the lesion can lead to a relapse of disease and a pathological fracture of the mandible could occur if the lesion interests a great vertical portion of the mandible, as in this patient. To date, the final result has been satisfactory but further follow-ups will be needed. This is in line with the literature.

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