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CASE REPORT

Eosinophilic Granuloma of Mandibular Condyle: Resection and Complete Regeneration

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Abstract Eosinophilic granuloma is a rare disease of abnormal proliferation of Langerhan cells affecting the jaws of young individuals. We report a case of complete regeneration of condyle in a 13 year old boy with eosinophilic granuloma condyle, who had undergone condylectomy.

Eosinophilic granuloma · Condyle · Regeneration

Introduction

Langerhan cell histiocytosis (LCH) is a rare disease in the head and neck region, characterized by abnormal proliferation of Langerhan cells. About 7 % of bony lesions occur in jaws, often seen in children, teenagers, or young adults. Bony lesions in the young age group can occur in any part of the skeleton, but soft tissue and particularly lung lesions occur between the ages of 20–40 years [1].

Jaw lesions may present with the classic picture of alveolar bone loss and mobile teeth, producing the "floating tooth" picture. LCH may mimic odontogenic neoplasm, sarcoma or osteomyelitis. Some may even present as multilocular radiographic appearance [1].

Case Report

A 13 year old male reported with complaint of pain and swelling in relation to the right preauricular region and difficulty in mouth opening. Clinical examination revealed a tender bony hard swelling at preauricular region. Mouth opening was restricted and ipsilateral submandibular lymph nodes were palpable.

Plain and contrast computed tomography revealed a lytic lesion with irregular margins in the right condyle extending to the subcondylar region causing break in the bony continuity from one cortex to the other with soft tissue component showing enhancement, particularly in the periphery (Fig. 1a–c). Fine needle aspiration cytology was inconclusive.

Under general anesthesia and nasoendotracheal intubation, right mandibular condylectomy was performed (Figs. 2, 3). Postoperative period was uneventful.

Microscopic examination of specimen revealed fibrocollagenous tissue and degenerating bony trabeculae. There were sheets of lymphocytes, plasma cells, polymorphs and plenty of eosinophils. On the basis of clinical, radiographic and histopathologic findings a diagnosis of eosinophilic granuloma (Langerhan's cell histiocytosis) of condyle was made (Fig. 4).

Patient was under regular follow-up for a period of 6 months without any significant postoperative complications. After that period patient did not turn up for further evaluation, though he was instructed to be under regular follow-up. Patient reported after a period of 6 years for routine examination. He had no complaints but radiographic evaluation revealed a completely regenerated right condyle without any pathology (Fig. 5).

Discussion

Langerhans cell histiocytosis (LCH) (previously known as Histiocytosis X) is characterized by intense and abnormal proliferation of Langerhans cells (bone marrow derived

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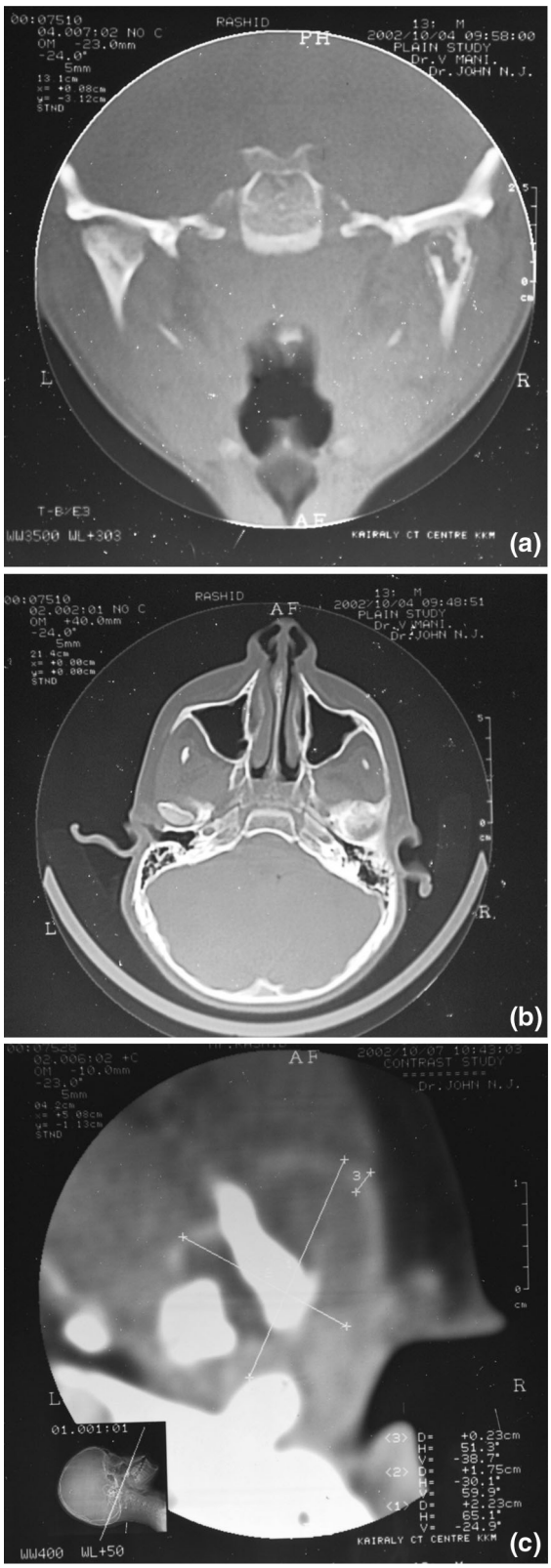

Fig. 1 **a**, **b** Preoperative CT, **c** contrast preoperative CT

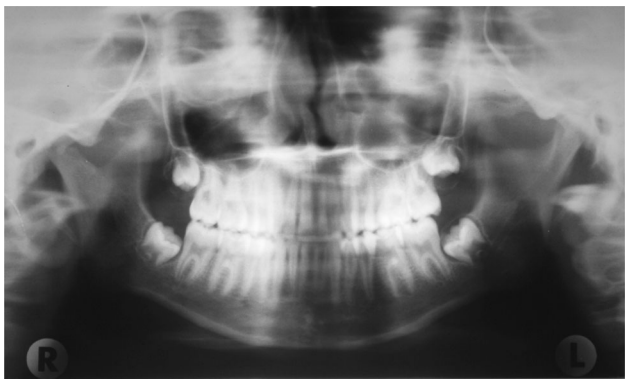

Fig. 2 Preoperative OPG

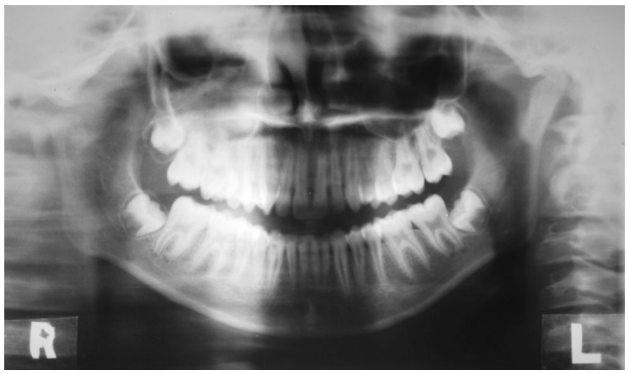

Fig. 3 Immediate postoperative OPG

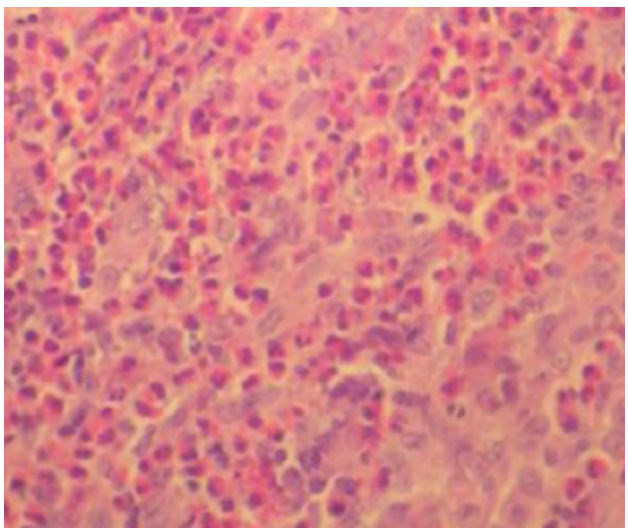

Fig. 4 Histopathology

histiocytes) along with variable number of leucocytes, eosinophils, neutrophils, lymphocytes, plasma cells and giant multi-nucleated cells causing tissue destruction [2–4].

Lichtenstein [5] gave a common terminology Histiocyttosis X for the triad of eosinophilic granuloma, Hand-Schuller-Christian, and Letterer-Siwe disease. In 1985 Feldmann et al. used the term Langerhans cell histiocytosis (LCH) to encompass the entities formerly called

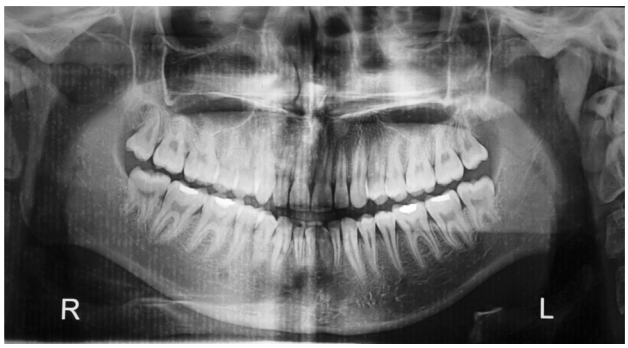

Fig. 5 6 year postoperative OPG

Histiocytosis X. Eosinophilic granuloma was described by Lichtenstein and Jaffe in 1940 [6]. Lichtenstein classified LCH into three clinical forms depending on the age of the patient when the lesions first appear and their distribution.

1. Chronic focal LCH (eosinophilic granuloma): It is the most frequent and benign form of LCH. It can appear as unifocal or multi focal lesion in a single, or multiple bones, with or without soft tissue, without systemic involvement and can present at any age [4].
2. Chronic diffuse LCH (Hand-Schuller-Christian disease): Most commonly seen in children and young adults. Clinically exophthalmos, osteolytic lesions of the cranium and diabetes insipidus forms the characteristic triad. Petechiae, purpura, ulcerations, lesions mimicking seborrheic dermatitis, pulmonary dysfunction, tachypnea, dyspnea and cyanosis, may appear [4, 7].
3. Acute disseminated LCH (Letterer-Siwe disease):This form of LCH follows a fatal form in short duration of time and children under 3 years of age are affected. Multiple organ systems are affected such as liver, lung, lymph nodes, skin, bone marrow.

Clinically may present as eczema, hepatosplenomegaly, otitis media, anemia, hemorrhages, lymphadenopathies and osteolytic lesions [4, 8].

Hashimoto-Pritzker syndrome (congenital reticulohistiocytosis): It is a purely cutaneous form of LCH, clinically manifesting as dark nodules on the trunk, face and scalp. The mucosae are always involved, without implication of other organs [9].

Eden [10], in 1998, staged LCH as:

Stage I—Single lytic bone lesion
Stage II—Multiple lytic bone lesions (both formerly termed as eosinophilic granulomata)
Stage IIIA—Bone plus soft tissue lesions, often associated with diabetes insipidus (pituitary involvement) or exophthalmoses (previously termed Hand-Schuller-Christian triad)
Stage IIIB—Soft tissue only, disseminated form (previously termed Letterer-Siwe disease)

Variable hypothesis have been proposed for the possible etiology of LCH. Immune system dysfunction results in hypersensitivity reaction to an unknown antigen and activation of histiocytes macrophage system [11, 12]. In advanced form of LCH deficiency of suppressor lymphocytes T_8 [11], autoantibodies, altered immunoglobins, abnormal lymphocytic response to mitogens and structural changes in the thymus have been found [13]. Viruses, bacteria, and genetic influences [14–16] interleukin-3, and tumor necrosis factor-alpha, tumor suppressor genes (p53), oncogenes (c-myc, h-ras), growth factors, cell surface immunologic markers and apoptotic factors have also been considered as etiologic factors in LCH as well [17]. LCH may manifest in the oral cavity first or oral lesion may be the only sign seen in the course of the disease [18, 19].

Most commonly bones are involved in all three forms of LCH. Mandible, maxilla, cranium are the most commonly involved and usually infiltration is seen. In mandible posterior body and ramus is more commonly involved [20]. Unifocal LCH (eosinophilic granuloma) is very rarely seen in the condyle as reported in our case. Patient may complain of pain and swelling in the region of lesion, mouth opening may be limited in case of condyle involvement as seen in our case. Mucosal [21, 22], periodontal manifestation [22–25], skin involvement [26] and cervical lymph adenopathy [19] also occurs in LCH.

Histopathology often suggests an inflammatory process, characterized, by the proliferation of Langerhans cells, which have abundant ill-defined cytoplasm and oval or indented nuclei that often have a central groove giving it a coffee bean appearance. Inflammatory cells lymphocytes and neutrophils are present, but eosinophils predominate. There may be infiltration of overlying epithelium. Giant cells and necrosis may be present, but mitoses are very uncommon. Definitive diagnosis is achieved by light microscopy and positivity for the CD1a antigen by immunostaining [1].

Treatment of eosinophilic granuloma is by surgical curettage, excision, or radiation therapy [27–33]. Irrespective of the treatment, the recurrence rate is low [33]. However, patient should be closely followed-up for an extended period.

We did right condylectomy in our case and patient was under regular follow-up for a period of 6 months. During that period he had no significant postoperative problems. Patient reported back to us after a period of 6 years and radiographic evaluation showed a completely regenerated condyle without any recurrence. He had no secondary deformity due to the condylectomy. Complete regeneration

of large portions of mandible in young individuals has been reported in literature [34–38].

The regeneration of condyle in our case may be attributable to the regenerative capability due to the higher cellular activity and availability of abundant mesenchymal cells in children to form osteogenic tissues.

Eosinophilic granuloma is a rare disease of the jaws mostly affecting young individuals which can be effectively treated by surgical excision. Patients should be kept under long term follow-up even though the recurrence rates are low.

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