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Langerhans Cell Histiocytosis in a 5-Year-Old Girl: A Case Report and Review of Literature

Abstract

Langerhans cell histiocytosis (LCH) is an uncommon hematological disorder affecting infants and young children. LCH is a rare disorder of the reticuloendothelial system associated with proliferation of Langerhans cells and mature eosinophils. LCH can involve any bone, but the most common are pelvis, ribs, skull, long bones, vertebra, and facial bones. In the skull, frontal and parietal bones are commonly involved followed by the jaws, where mandible is more commonly affected than the maxilla. In this article, we report a case of LCH in a 5-year-old child involving the mandible. Swelling of one side of the face and aggressive periosteal reaction led to the diagnosis of monostotic LCH. The manuscript also summarizes the results of a literature search in PubMed of reported cases of LCH over the past 10 years.

Keywords: *Langerhans cell histiocytosis, mandible, periostitis*

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Introduction

Langerhans cell histiocytosis (LCH) is an uncommon hematological disorder affecting infants and young children.[1] LCH, previously termed as histiocytosis X, is characterized by an uncontrolled stimulation and proliferation of normal antigen presenting cells, Langerhans cells, and mature eosinophils.[2] The term histiocytosis refers to the proliferation of histiocytes and other inflammatory cells, and "X" was added to denote the unknown cause of the disease. The accumulation of the pathological Langerhans cells causes infiltration and destruction of the local tissues.[3] The incidence of LCH is relatively low with an estimation of 2–5 cases per million per year.[1]

LCH mainly comprises of three morphologically similar lesions, namely eosinophilic granuloma, Hand–Schuller–Christian syndrome, and Letterer–Siwe syndrome classified by Lichtenstein in the year 1953.[3,4] The cause of this disease is not clearly known; however, there is evidence that the disorder is a presentation of an immunological variation.[5] In this article, we report a case of LCH involving the mandible with an overt swelling of one side of the face which was diagnosed with the help of imaging modalities and histopathological investigation.

Case Report

A 5-year-old girl reported to the department of oral medicine and radiology with a chief complaint of swelling in the left side of the face since 1 and ½ months. The patient complained of pain in the same region during mouth opening and on chewing. The patient's parents gave a history of consulting local doctor for the same where antibiotics and analgesics were prescribed. Swelling persisted even after medications.

On extraoral examination, a solitary diffuse swelling was noticed on the left side of the face measuring around 6 cm × 5 cm in size extending superoinferiorly from the ala-tragal line to the inferior border of the mandible, mediolaterally from the left commissure of the mouth to the preauricular area [Figure 1a]. Swelling was firm in consistency and tender on palpation. Local rise in temperature was present. Solitary right and left submandibular lymph nodes were palpable, firm in consistency, mobile, and nontender. On intraoral examination, obliteration of the buccal vestibule was noted in the left posterior mandibular region [Figure 1b]. On palpation, tenderness was present in the distal aspect of the deciduous left mandibular second molar region and ascending body of

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the ramus region. Expansion and perforation was present in the medial and lateral aspect of the ascending body of the ramus of the mandible. Laboratory investigation was within normal limits.

Panoramic imaging showed an ill-defined osteolytic lesion extending from the distal aspect of tooth bud of mandibular left permanent second molar till the coronoid process and sigmoid notch of the mandible [Figure 2]. Computed tomography (CT) scan of the patient showed areas of ill-defined destruction and pathologic fracture of the ramus of the mandible. Aggressive periosteal reaction was noted giving a sun ray-like appearance [Figure 3].

Based on the age, rapidly aggressive nature, clinical presentation, and radiological features, a differential diagnosis of LCH, Ewing's sarcoma, and nonsuppurative osteomyelitis was considered.

Histopathological examination of incisional biopsy specimen showed extensive proliferation of histiocytes with indistinct cytoplasmic borders and rounded vesicular nuclei with interspersed eosinophils suggestive of LCH [Figure 4a]. Immunohistochemistry showed positivity for CD68, CD1a, and S-100 confirming the diagnosis [Figure 4b and c]. Positron emission tomography revealed no active lesion elsewhere in the body.

The patient was treated with surgical corticotomy under general anesthesia of the affected side with extraction of the left mandibular second and third molar tooth buds. The patient is currently under periodic follow-up.

Discussion

Histiocytosis is a term that refers to a group of rare disorders of the reticuloendothelial system. LCH is associated with proliferation of specialized bone marrow-derived antigen presenting dendritic cells, namely the Langerhans cells and mature eosinophils.[2] Since the proliferating histiocytes involved in LCH are phenotypically similar to that of the Langerhans cells found in the normal mucosa and skin, the condition was named as LCH.[3,6]

Various hypotheses have been proposed explaining the etiology of LCH. The presence of inclusion bodies resembling the Birbeck granules of Langerhans cells classifies LCH as a proliferative disorder of the Langerhans cells or their marrow derivatives.[4] Studies have also shown that LCH is a reactive disease that may result from environmental or other triggering factors, which leads to the aberrant reaction between Langerhans cell and T-lymphocytes.[7]

LCH is more frequent in males than in females with a ratio ranging from 1.1:1 to 4:1.[4] It predominantly affects children and young adults.[4-6] The present case was reported in a 5-year-old girl.

Eosinophilic granuloma of the monostatic and multifocal form is a type of LCH which affects children and young adults and manifests as solitary or multiple skeletal lesions without extraskeletal involvement. The chronic and disseminated form of LCH termed as the Hand–Schuller–Christian syndrome consists of skeletal and extraskeletal lesions usually affecting children

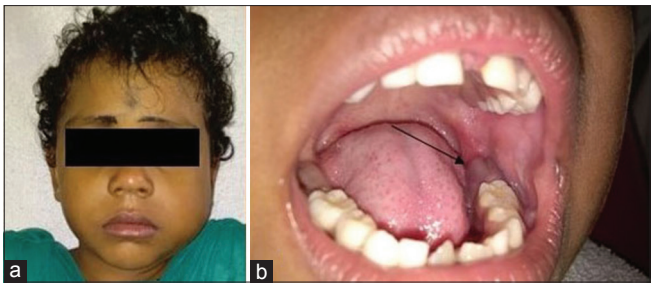

Figure 1: (a) Extraoral swelling on the middle and lower one-third of the face right side. (b) Intraoral obliteration of the buccal vestibule was noted in the left posterior mandibular region

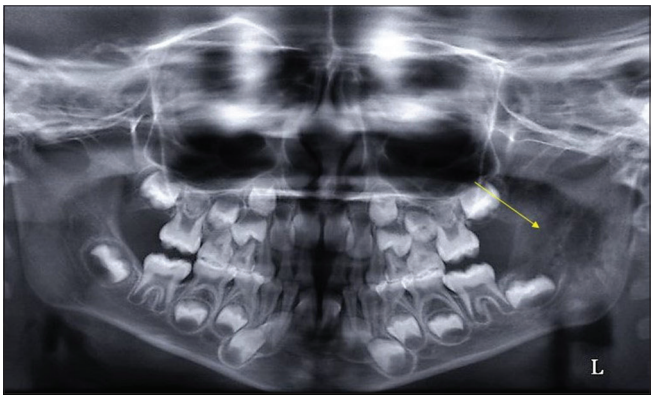

Figure 2: Panoramic imaging showed an ill-defined osteolytic lesion extending from the distal aspect of tooth bud of mandibular left permanent second molar till the coronoid process and sigmoid notch of the mandible

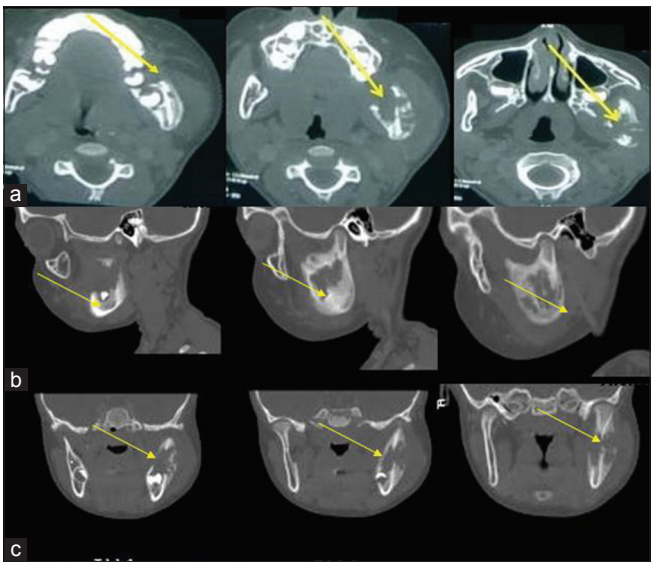

Figure 3: (a) Axial, (b) Sagittal and (c) Coronal sections of computed tomography scan showing areas of ill-defined destruction and pathologic fracture of the ramus of the mandible. Also aggressive periosteal reaction can be noted giving a sun ray-like appearance

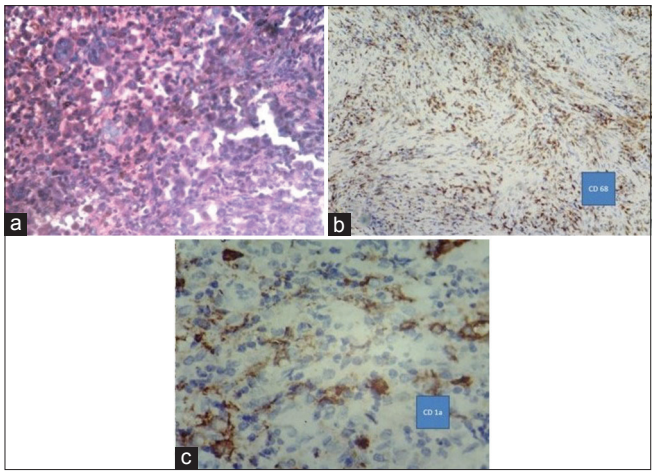

Figure 4: (a) Histopathological picture showing extensive proliferation of histiocytes with indistinct cytoplasmic borders and rounded vesicular nuclei with interspersed eosinophils suggestive of Langerhans cell histiocytosis . (b and c) Immunohistochemistry showed positivity for CD68, CD1a, and S-100 confirming the diagnosis

below 3 years of age. The Letterer–Siwe syndrome is a disseminated acute or subacute form of LCH, that is, most often fatal because of extensive skeletal and extraskeletal lesions; this form usually affects infants and children below 3 years of age.[3,4] The present case represents monostotic eosinophilic granuloma of the mandible.

The relative incidence of organ system involvement in LCH is as follows: bone in 80% of the cases; skin 60% of the cases; liver, spleen, and lymph nodes 33%; lungs and orbit in around 25% of the cases; and maxillofacial in around 25% of the cases.[1] Skeletal involvement can involve any bone, but the most common are pelvis, ribs, skull, long bones, vertebra, and facial bones.[4] In the skull, frontal and parietal bones are commonly involved followed by the jaws.[7,8] Mandible is more commonly involved when compared to the maxilla.[2] In the present case, only the posterior aspect of the mandible was involved.

A literature search for case reports consisting of the following words "'Langerhans cell histiocytosis of the jaw" and "eosinophilic granuloma of the jaw" for the past 10 years was carried out in PubMed database which reported 38 cases of LCH of the jaw. It consisted of 12 cases below the age of 10 years, 6 cases between the age groups of 11–20 years, and 13 cases above the age group of 20 years. Out of the 38 cases, information regarding the age for 7 cases were unavailable. Only mandible was involved in 25 [68%] cases, maxilla alone was involved in 6 (15.7%) cases, and both maxilla and mandible were involved in 4 (10.52%) cases, and in 3 cases, the site involved was not specified [Table 1].

In a review of 1120 patients, oral involvement was reported in around 10% of the cases.[43] Intraorally, it usually appears as a soft-tissue swelling or ulceration of the gingiva.[44] The most common site is the mandibular molar area, followed by the ramus region.[2] In the present case, obliteration of the buccal vestibule with no surface ulceration, discharge, or bony exposure was noticed.

Destruction of the lamina dura, floating teeth appearance indicative of destruction of alveolar bone is evident on radiographs.[2] Other radiographic features include localized, punched-out radiolucencies with no evidence of calcification and sclerosis.[4] Mandibular lesions are ill-defined with single or multiple punched-out radiolucencies without corticated rimming suggestive of active disease.[1] Radiographically, eosinophilic granuloma typically presents as punched-out osteolytic lesions with or without periosteal reaction.[45] These radiological features are present due to the destruction caused by the Langerhans cells which may cause pathological fracture of the bone.[1] In the present case, it manifested as an osteolytic ill-defined lesion with pathological fracture and periosteal reaction.

Based on the age, rapidly aggressive nature, clinical presentation, and radiological features, a differential diagnosis of Ewing's sarcoma, LCH, and nonsuppurative osteomyelitis was considered. Both Ewing's sarcoma and LCH show similar radiological appearance. Ewing's sarcoma usually affects long bone and very rarely affects the mandible. The other possible diagnosis is nonsuppurative osteomyelitis based on the history of trauma, nature of the lesion, and moth-eaten appearance of the mandibular ramus area noticed in the CT scan.

Histopathologically, LCH presents as a diffuse infiltration of pale-staining mononuclear cells that resemble histiocytes with indistinct cytoplasmic borders and rounded vesicular nuclei. Multiple eosinophils can be seen typically interspersed among the histiocytes, plasma cells, lymphocytes, and multinucleated giant cells.[46] In the present case, similar histopathological features were noticed.

On immunohistochemistry, LCH is positive for S-100 and anti-CD-1a, which was also seen in the present case.[6] LCH is characterized by antigen Ki-67 that selectively binds to a nuclear antigen which is only expressed by proliferating cells.[6]

Treatment modalities available for LCH depend on the site of the lesion, its extent, and the number of lesions present. Depending on this either surgical curettage, radiotherapy or chemotherapy can be used alone or in combination.[47,48] In the present case, as the lesion was unifocal and involved only mandible, surgical curettage was carried out. However, if there are multifocal involvement and associated systemic disease, chemotherapy should be considered. Recurrence rates around 1.6% to 25% are noticed, and a close and regular follow-up for a long period is advised.[4]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

Table 1: Clinical characteristics (age and site) of cases of LCH of jaw reported in PubMed in the last 10 years

Author name	Number of cases reported	Age	Site
Varsha *et al*. 2016[9]	1	Child aged 4 year	Hard palate
Vargas *et al*. 2016[10]	1	Adolescent aged 16 years	Maxilla
Fiorini *et al*. 2016[11]	1	*	Mandible
Xian *et al*. 2015[12]	1	Adult patient	Mandibular condyle
Emmanouil *et al*. 2015[13]	1	Child aged 8 years	Hand–Schuller–Christian syndrome
Vennamaneni *et al*. 2015[14]	1	Adult aged 29 years	Mandible
Sherwani *et al*. 2014[15]	1	Child aged 4 years	Mandible
Liu *et al*. 2014[16]	1	Infant aged 21 months	Hard palate
Zajko 2013[17]	1	Adult aged 45 years	Mandible
Felstead *et al*. 2013[18]	1	*	Mandible
Alshadwi *et al*. 2013[19]	1	Infant	Mandible
Terada *et al*. 2013[20]	1	Adult aged 46 years	Maxilla and mandible
Lee and Yoon 2012[21]	1	*	Two cases reported in mandible
Ge *et al*. 2012[22]	1	Adult aged 34 years	Mandible
Yepes *et al*. 2012[23]	1	Adult aged 23 years	Mandibular condyle
Lajolo *et al*. 2012[24]	2	a. Adult aged 71 years b. Adult aged 77 years	a. Mandible b. Hard palate
Shetty *et al*. 2012[25]	1	Child aged 14 years	Bilateral maxilla
Aydin *et al*. 2012[26]	1	Adult aged 45 years	Mandible
Anastasilakis *et al*. 2012[27]	1	Adult aged 21 years	Mandible
Martin *et al*. 2011[28]	1	Infant aged 4 months	Mandible
Murray *et al*. 2011[29]	1	Infant aged 22 months	Maxilla, mandible, and hard palate
Bas *et al*. 2011[30]	1	Child aged 7 years	Mandible
Dholam *et al*. 2011[31]	1	Child aged 12 years	Mandible
Alloh Amichia *et al*. 2010[32]	1	*	Jaw
Muramatsu *et al*. 2010[33]	1	Infant aged 13 months	Mandible
Park and Chung 2010[34]	1	Child aged 1 year	Mandibular condyle
Gaundong Mbéthé *et al*. 2010[35]	2	*	Jaw
Altug *et al*. 2010[36]	1	Adult aged 20 years	Mandible
Esen *et al*. 2010[37]	1	*	Mandible
Bouw and Nout 2009[38]	1	Infant aged 6 months	Maxilla
Rees and Paterson 2009[39]	1	Adult aged 31 years	Jaw
Helbling-sieder *et al*. 2009[40]	1	Adult aged 27 years	Mandible
Guiglia *et al*. 2009[41]	1	Child aged 13 years	Mandible
Orzechowska-Wylegała *et al*. 2008[42]	2	a. Adult aged 28 years b. Child aged 2 years	a. Mandible b. Maxilla, mandible

*Details could not be accessed or were not available

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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