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Mandibular LCH Masquerading as Parotid Enlargement: A Diagnostic Challenge

Case Report

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Abstract

Langerhans Cell Histiocytosis (LCH) is an uncommon disorder marked by the proliferation of specialized dendritic cells that can infiltrate various organs. Characterized by its diverse clinical presentations, LCH most frequently appears as solitary eosinophilic granulomas in bone, primarily affecting children. Despite its rarity, with an incidence ranging from 2 to 5 cases per million annually, LCH can present at any age and often mimics other conditions, making accurate diagnosis challenging. Clinical manifestations may vary widely, from localized bone lesions to systemic involvement.

In pediatric patients, LCH can present as unifocal bone lesions, often leading to misdiagnosis if not thoroughly evaluated. A case in point is an 18-month-old female who presented with a progressive swelling in the left cheek, initially suspected to be a dental infection. Subsequent imaging and biopsy confirmed the diagnosis of unifocal LCH of the mandible. This case highlights the importance of considering LCH in differential diagnosis, especially when dealing with atypical bone lesions in children.

Keywords: Langerhans Cell Histiocytosis; Eosinophilic Granuloma; Mandibular Lesion; Pediatric Bone Disease;Ameloblastoma.

Introduction

Langerhans cell histiocytosis (LCH) is a rare systemic disorder characterized by idiopathic proliferation of histiocytes, called Langerhans cells, in different organs including the bones, lungs, central nervous system, liver and spleen, skin, thymus and lymph nodes. The severity and clinical behaviour depend on the number and type of organ systems involved. Skeletal involvement is common and may affect one or multiple bones. Involvement of a solitary bone was previously referred to as eosinophilic granuloma (EG) and is the most common presentation of LCH in children.[1]LCH is not common disease with reported incidence of 0.2 – 2.0 cases per one lakh children under fifteen years old.[2] Clinical presentations of LCH can range from localized bone lesions to severe multisystem involvement, which can be potentially life-threatening. Diagnosing LCH requires a thorough approach that includes clinical examination, histopathological studies, immunohistochemical tests, and imaging techniques. Plain radiography, computed tomography and magnetic resonance imaging are the most used techniques for detection and characterization of the lesion. While the outlook is generally positive, especially for children, there remains a significant risk of relapse and

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complications, particularly in cases with multisystem involvement. This article details the case of an 18-month-old female child diagnosed with unifocal bony Langerhans Cell Histiocytosis (LCH), specifically affecting the mandible.

Case Presentation

We present a case of Eighteen months old girl who was brought to our hospital with a rapidly growing swelling on her left cheek that had been present for more than three weeks. She had a fever one week earlier but had not experienced any weight loss or reduced appetite. On physical examination, a hard non-tender swelling was noted on her left cheek. Suspecting a potential parotid gland pathology, the clinician recommended an ultrasound of the neck.Ultrasound revealed a well-circumscribed, expansile mass in the body of left mandible with low echogenicity and internal vascularity. The lesion caused significant destruction of both the cortical and medullary bone surfaces of the mandible (Figure 1a)and (Figure1b). Suspecting mandibular pathology, further imaging was advised. The CT imaging revealed a large, well-defined expansile lytic lesion involving the ramus and mandibular condyle on the left side, with extension to the TM joint and associated cortical discontinuity(Figure2a) (Figure2b) and (Figure2c). To know the soft tissue extension, MRI neck with contrast was done which revealed ill-defined, expansile altered signal intensity lesion in the left mandible, extending from the angle of the mandible to the condylar and coronoid processes. The lesion is associated with significant cortical thinning and was in close proximity to the masseter muscle, with a discernible loss of the fat plane separating the lesion from the adjacent muscle.Post-contrast imaging shows subtle enhancement of the lesion. The overlying muscle of mastication shows T2 hyperintensity with thickening and enhancement, consistent with inflammation or infiltration. The parotid gland is separately visualized and does not appear to be involved by the lesion (Figure 3a-3d). Differential Diagnosis considered were Langerhans Cell Histiocytosis (LCH), osteomyelitis, Ewings sarcoma and Ameloblastoma .

Ultrasound guided biopsy was done(Figure 4). Samples were sent for HPE examination.Histological features were s/o Langerhan cell histiocytosis and subsequent IHC was positive for S100 and anti-CD-1a (Figure 5).

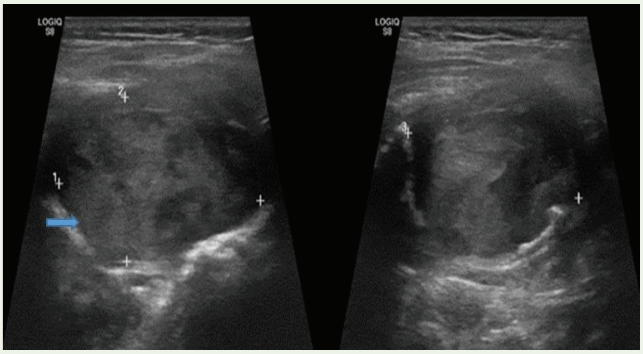

Figure 1: Sagittal and axial ultrasound image showing a well-circumscribed, expansile lesion in the body of left mandible with low echogenicity and internal vascularity having significant destruction of both the cortical and medullary bone surfaces of the mandible.

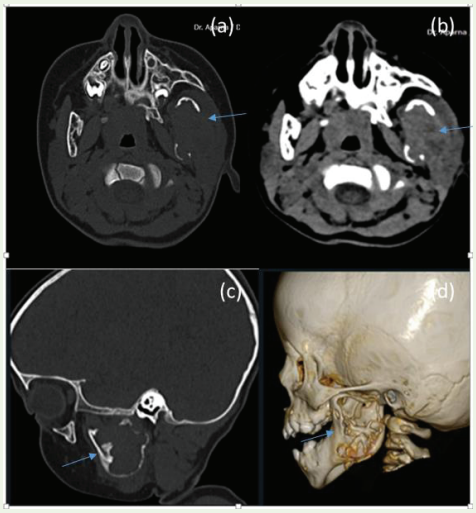

Figure 2: CT axial, sagittal and reformatted images in bone and soft tissue window showing a large, well-defined expansile lytic lesion involving the ramus and mandibular condyle on the left side, with extension to the TM joint and associated cortical discontinuity.

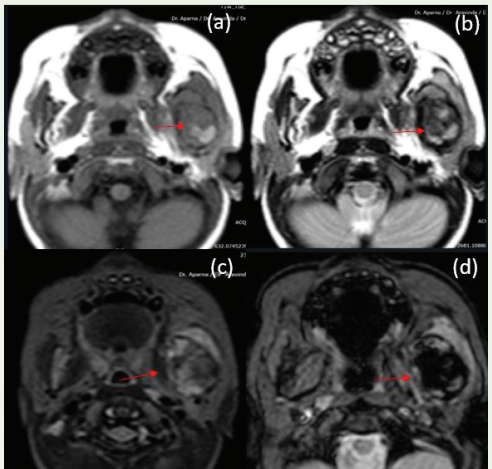

Figure 3: MRI neck with contrast T1 (a), T2 (b), T1 fat sat contrast(c) and GRE (d) axial images showing ill-defined, expansile altered signal intensity lesion in the left mandible, extending from the angle of the mandible to the condylar and coronoid processes. Post-contrast imaging shows subtle enhancement of the lesion.

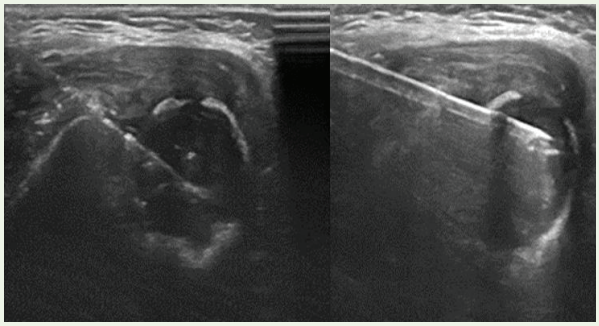

Figure 4: Ultrasound guided biopsy images of the mandibular lesion.

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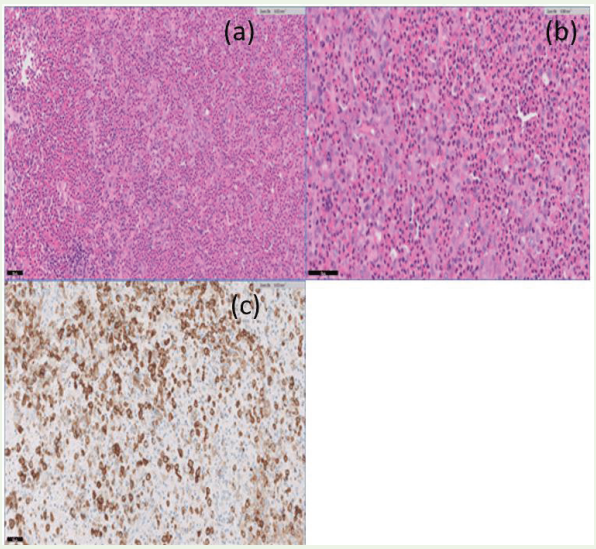

Figure 5: Histopathological images in low power (a) and high power (b) showing extensive proliferation of histiocytes with indistinct cytoplasmic borders and rounded vesicular nuclei with interspersed eosinophils suggestive of Langerhans cell histiocytosis. Immunohistochemistry (c) showed positivity for CD1a confirming the diagnosis.

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.[3] 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. Skeletal involvement 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.[3] Mandible is more commonly involved when compared to the maxilla [4]In the present case, only the posterior aspect of the mandible was involved. While the condition is uncommon, its presentation in the mandible aligns with the literature indicating that the mandible is the second most common site of osseous involvement in LCH, following the calvarium.[1]

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 fever, nature of the lesion, and moth-eaten appearance of the mandibular ramus area noticed in the CT scan. Imaging plays important role in diagnosing bone lesions, particularly CT and MRI where we can know the characteristics, extent and adjacent soft tissue involvement which will help in narrowing down the differentials and in management.[5]

Histopathological analysis is critical for accurate diagnosis. 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.[6] In our case, similar histopathological features were noticed with IHC positive for S-100 and anti-CD-1a [6]LCH is characterized by antigen Ki-67 that selectively binds to a nuclear antigen which is only expressed by proliferating cells. [7] These characteristic features help differentiate LCH from other conditions with similar radiological presentations.

Treatment strategies for LCH vary based on disease extent and location. For isolated bone lesions, conservative approaches such as surgical curettage and local steroid injections are often effective. In contrast, multisystem disease may require more intensive treatments, including systemic chemotherapy, radiotherapy, and/or surgery[8,9].

Conclusion

Unifocal LCH of mandible in an infant is a rare condition with a reported incidence of 0.2 - 2.0 cases per 1,00,000 children under 15 years old, but an important diagnosis to consider when evaluating mandibular lesions in children. The clinical presentation, radiological features, and histopathological findings are crucial for distinguishing it from other pathologies. Early diagnosis and prompt treatment are essential to prevent complications such as extensive bone damage and facial disfigurement. Regular follow-up and a comprehensive treatment approach are necessary to manage the condition effectively and ensure favourable outcomes.

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