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Case Report
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Langerhans Cell Histiocytosis of Left Lower Gingivo Buccal Groove in Adult-Monosite, Single System and Role of Local Radiotherapy - A Case Report

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Abstract

Langerhans Cell Histiocytosis (LCH) is a very rare benign disease. It was previously known as Histiocytosis 'X'. Histologically it is characterized by the atypical clonal proliferation of immature Langerhans cell histiocytes in addition to eosinophils, neutrophils and plasma cells. We report a case of Langerhans cell histiocytosis of left lower buccal groove in a 51-year-old woman. She was treated with local radiotherapy. This case is being reported due to rarity of LCH in adults.

Keywords: Langerhans cell histiocytosis (LCH); Radiotherapy; Monosite; PET-CT

Introduction

Langerhans Cell Histiocytosis (LCH) is a rare benign disease with an incidence of 0.1 per one lakh population in adults and 0.5 to 2 per one lakh population in children. In adults it usually occurs in the 4th to 5th decade of life. LCH can present as a single system or multisystem disease with unifocal or multifocal involvement. The most common organ involved in adults is lung followed by bone and skin. The most common site in children is bone followed by skin, reticuloendothelial system, bone marrow and orodental region [1-4].

We report the rare presentation of LCH in a middle-aged woman. The site of the lesion was left lower buccal groove, which is a rare site in adults.

Case Presentation

This 51-year-old middle aged woman a known type-2 diabetic presented with the complaints of pain and ulcer in left lower buccal groove of three months duration. Initially she consulted a dentist and underwent first molar tooth extraction and biopsy of the lesion. Biopsy was consistent with Langerhans cell histiocytosis. She was referred for local radiotherapy in view of monosite single system LCH. Clinical examination revealed ulcerative lesion measuring approximately 3 cm × 2 cm in the left lower buccal groove extending from first premolar to 2nd molar tooth (Figure 1). Rest of the oral cavity was normal. No significant lymphadenopathy or organomegaly. Her complete blood counts, liver and renal function tests were within normal limits. Viral markers (HIV, HCV, HBV) were negative. Blood sugar was under control. Biopsy of the lesion done in our institute revealed hyperplastic squamous epithelium with down growths. Sub-epithelium showed dense infiltrate of lymphocytes and plasma cells and focal areas showed sheets of histiocytes with oval grooved nuclei admixed with numerous eosinophils. Some of fragments showed mucosal ulceration and necrosis (Figure 5). Immunohistochemistry (IHC) was positive for CD1a, CD45, CD68, S100 protein and Ki 67 (Figure 6). PET-CT (Positron Emission Tomography) scan showed destructive lytic lesion in the left side of ramus of mandible in the first molar region with soft tissue involvement (SUVmax-10.1) extending into the adjacent left lower buccal space. There is no extension into the retro-molar trigone. There is no involvement of skin. There is no hepatosplenomegaly or lymphadenopathy (Figure 2).

This patient was grouped under Group 3 single system monosite low risk organ as per histiocyte society international protocol LCH III trial and was started on local radiotherapy. The patient received a total dose of 20 Gy to the clinical target volume, 2 Gy per fraction, using 6 MV photons after taking informed consent. Three-dimensional conformal radiotherapy technique was used. The

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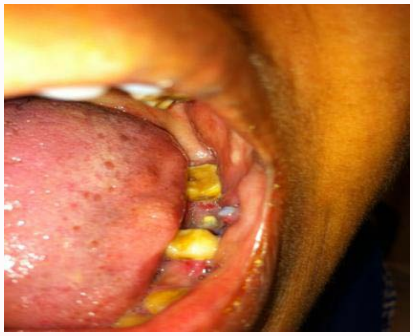

Figure 1: Image showing ulcerated lesion (LCH) in left lower buccal groove.

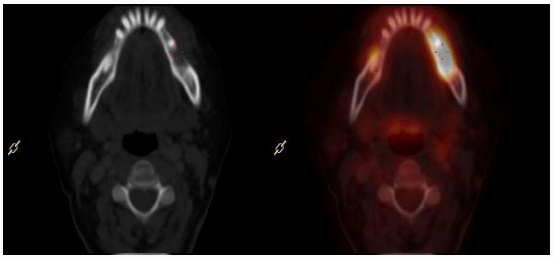

Figure 2: Pre-treatment PET CT-scan images showing lytic lesion in left side of mandible in the first molar tooth region.

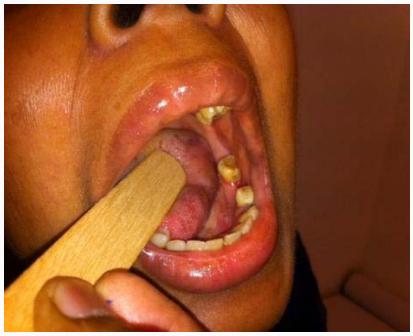

Figure 3: Image taken at follow up after 4 months showing complete regression of the lesion.

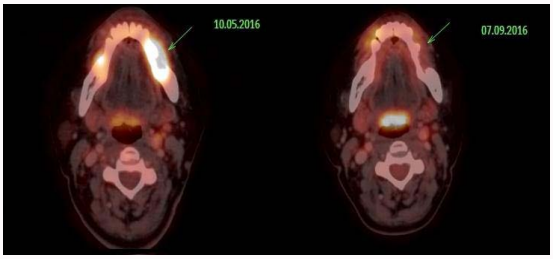

Figure 4: PET–CT scan done 4 months after treatment showing complete regression of the lesion.

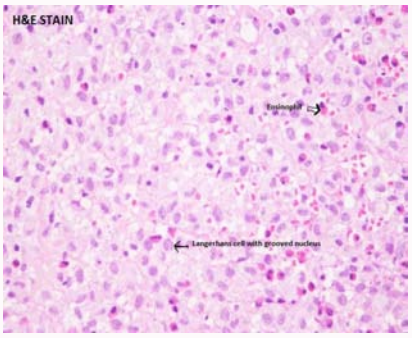

Figure 5: Image showing H&E stain – Langerhans cell with grooved nucleus, Eosinophils, Neutrophils, Plasma cells and lymphocytes.

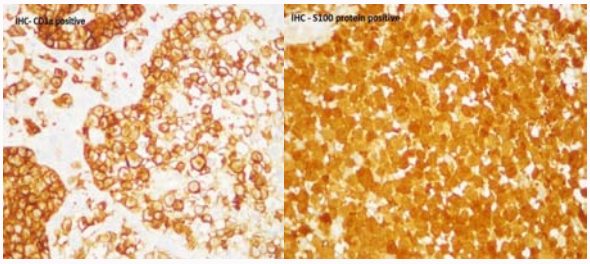

Figure 6: Image showing Immunohistochemistry (IHC) stains – CD1a and S100 protein.

clinical target volume included the gross tumor with a 1 cm margin. She had satisfactory pain relief during radiotherapy.

Four months after completion of treatment, she is asymptomatic. Clinical examination and PET-CT scan showed complete regression of the lesion (Figure 3 and 4). Patient was disease free after 4 years of treatment.

Discussion

LCH is very rare in adults. Lung is the most common site of involvement in adults followed by bone and skin. In LCH the most commonly involved bony sites are mandible in adults and skull bone in children. Genital and oral mucosal involvements are common in adults compared to children. Radiological investigations aid in identifying the site of lesion and for assessing single or multisystem involvement. Under light microscopy, histiocytosis is distinguished by immature dendritic cells with nuclear grooves and nuclear indentation with low nuclear to cytoplasmic ratio (Figure 5). Immunohistochemically, it can be identified by CD1a, CD207 and S100 protein positivity. CD1a and S100 were positive in this case [5]. Presence of Birbeck granules under electron microscopy is characteristic of LCH. Patients can be categorized into three groups based on system as well as risk organ involvement (lung, liver, spleen, bone marrow) as per histiocyte society International protocol LCH III trial. Group I include multisystem risk organ involvement (hematopoietic, liver, lungs or spleen), Group II includes multisystem without risk organ involvement and Group III includes single system multifocal bone disease or localized special site involvement (like orbit, temporal bone and mastoid) with intracranial soft tissue extension. Based on these categories, the patient was put under Group 3 because of single system mono site low risk organ involvement.

Patients with localized LCH involving bone, lymph node or skin will have a good prognosis and responds well to local treatment. Multifocal disease, multisystem involvement and localized special site involvement, such as “CNS-risk” lesions with intracranial soft tissue extension or vertebral lesions with intraspinal soft tissue extension are associated with high relapse rate and poor prognosis. These patients require systemic therapy with combination of other modalities [7].

Adult LCH is treated with surgery, radiotherapy, chemotherapy, steroids or combined modality therapy. In patients with localized

single skeletal lesions without any organ dysfunction, biopsy or curettage is the initial mode of treatment. Treatment options for single bone lesions that recur or remain symptomatic after surgery include non-steroidal anti-inflammatory agents or local steroid injections and local radiation therapy [7]. Survival and local control were found to be better in patients treated with surgery and radiotherapy when compared with single modality treatment [6].

Conclusion

This case is being reported due to rarity of LCH in adults. Radiotherapy is a good option for patients with monosite and single system disease. It is non-invasive, less morbid, cosmetically sound and patient compliance is also better.

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