

retention, distinctly audible phonetics, acceptable esthetics and good functional occlusion. The prosthesis used in these cases were implant as well as tissue supported as compare to the cantilever supported on bicortical implants in symphysis region hence masticatory efficiency was little higher. During review each patient was evaluated for fit, retention and stability, effect on perioral structures and masticatory experience along with psychological satisfaction with esthetics.

Stabilisation of the bone volume is of prime importance after the completion of the treatment. In our study, after complete removal of the distraction device, the increased bone volume was stable till the end of observation phase, which commensurate with the findings of Kelsper et al¹⁰. After six months of review clinically and radiographically, we have recorded stable conditions with no failure of the incorporated prosthesis. Radiologically, the distraction area seems to remodel as per the functional requirement, withstand masticatory forces transmitted through the implants.

We recommend a further long term study of at least 05 years to assess the long term status of the distracted bone, clinically and histologically. The main disadvantages of this study were the cost factors of various biomedical devices, technique sensitivity of the procedure and comparative multiple hospitalizations.

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Langerhan's Cell Histiocytosis: A Case Report and Review of Literature.

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Abstract: Langerhans cell histiocytosis (LCH) formerly known as histiocytosis X is one of a group of poorly understood disease of histiocytes. LCH previously has been considered a reactive polyclonal disease of immune regulation and not a true neoplasm. More recent evidence, however, has demonstrated clonal proliferation, a key neoplastic feature. The proliferation of dendritic cell, in addition to histiocytes, eosinophils, lymphocytes and plasma cells are the histological hallmark of LCH. Mandibular involvement associated with LCH is uncommon in young children. This article presents a case of LCH occurring in the mandible of a 9 year old male child which was managed conservatively.

INTRODUCTION

Langerhan cell histiocytosis is a relatively rare and unique disease process characterized by an abnormal proliferation of immature dendritic cells¹. The nomenclature – histiocytosis X was coined by Lichenstein in 1953 to account for 3 clinical varieties which showed some histological characteristics in common- Eosinophilic granuloma, Letterer-Siwe Syndrome and Hand Schuller Christian Syndrome.² The term 'histiocytosis' refers to proliferation of histiocytes and other inflammatory cells. The letters 'X' denote the unknown etiology of the disease². In 1973, LCH terminology was adopted due to the fact that histiocytes involved in the disease present

a phenotype which is similar to Langerhans cell found in normal mucosa and skin¹. The annual incidence of LCH is reported to be 0.5–5.4 million children per year³. It is predominantly a childhood disease and more than 50% of affected individuals are younger than 15 years. Oral lesions may be the earliest manifestation and with the aid of panoramic radiograph of the jaws these lesions will be detected at an early stage. The incidence of mandible being affected is twice as often as the maxilla⁴. The purpose of this article is to present a case report of conservative management of a 9 year old child affected with LCH of mandible and to enhance the understanding of LCH.

CASE REPORT

A 9 year old child reported to the department of dental and maxillofacial surgery with chief complaint of pain and swelling on right mandibular posterior region since 1 month. On extra oral examination facial asymmetry was seen on right lower facial region.

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The swelling was diffuse, approximately 3X2 cm in size, soft to firm in consistency, tender and extending from commissure of lip to anterior border of right Masseter muscle. On lymph node examination bilateral submandibular lymph nodes were enlarged, mobile, single and tender on palpation. On Intra oral Examination buccal sulcus was obliterated and mild bulge was noted buccally in the region of right lower deciduous molars (fig 1). On percussion right lower deciduous molars were painful and mobile. Remaining teeth and oral mucosa appeared unremarkable. OPG revealed (fig 2) a well defined lytic lesion around unerupted, developing premolars and canine in right body region of mandible. The lesion was extended from alveolar bone to inferior border of mandible superior-inferiorly and distal surface of developing canine to distal surface of second developing premolar antero-posteriorly. FNAC (fig 3) was done which showed highly polymorphous cell population, in which majority of cells were singly scattered, large, round to oval with pale staining nuclei, nuclear folding, grooving and sieve like chromatin. The cytoplasm was moderate and pale blue. Background showed plenty of eosinophils, few mature lymphocytes, giant cells, and macrophage showing phagocytosis(fig 3a,3b). These cytological features were suggestive of Langerhans Cell Histiocytosis. FNAC from submandibular lymph node showed Reactive Lymphoid Hyperplasia. CECT mandible (fig 4) showed a well defined radiolucent lesion with scalloping margins, 3x3x2cm in size involving developing premolar and canine in right side of mandible. Expansion of both cortical plates was noticed but no cortical breach or periosteal reaction was seen. Blood Investigations and x-ray of skull, femur was done to rule out systemic involvement.



Figure 1: Intra oral view showing vestibular swelling and mobile mandibular right primary first molar.



Figure 2: Panoramic radiograph of the patient depicts a well defined lytic lesion in right body region of mandible involving the permanent mandibular right primary first molar.

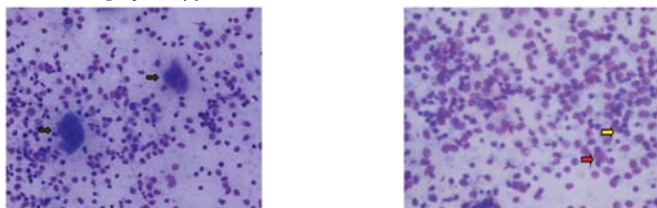


Figure 3: FNAC staining under GIEMSA 40X showing multinucleated giant cells (black arrows, figure:3a) and Yellow arrow- Eosinophils, Red arrow- Lymphocytes(fig:3b)



Figure 4: CT scan of 9 year old male that demonstrates right mandibular alveolus bone involvement with expansion of bucco-lingual cortical plates surrounding the primary first molar.

Incisional biopsy was done under local anesthesia from the lesion in the mandible. Grossly specimen was grey white to grey brown in color and hard in consistency. Microscopically sections showed tumor arranged in diffuse sheets. Individual tumor cells were round to oval in shape with eosinophilic cytoplasm, with nuclear grooving and indentation with 0-1 prominent nucleoli. Background showed presence of chronic inflammatory infiltrate comprising lymphocytes, plasma cells and eosinophils (fig5). On Immunohistochemical staining, CD1a and S-100 were positive (fig 6a, 6b). A definitive diagnosis of LCH was made.

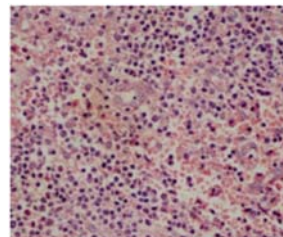


Figure 5: H & E 40x: Histiocytes with nuclear grooves, lymphocytes and eosinophils.

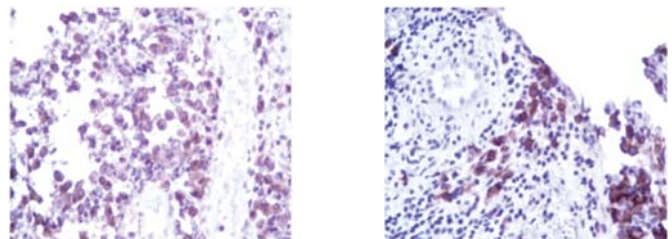


Figure 6: Gingival biopsy exemplifies the immuno-histochemical staining of LCH cells for S-100 (fig:6a) protein and CD1a (original magnification 25x in fig:6b).



Figure 7: Post op frontal view after 24 months of follow up with eruption of right mandibula premolars and canine



Figure 8: Post op panoramic radiograph of the patient after 24 months showing complete resolution of lesion.

In this presenting case considering the age of child, a conservative treatment management was done with intralesional injections of steroids (depo-medrol 40mg). A total of six injections, at interval of one week were given. Follow up was done at regular interval of 3 months up to 2 years; beginning of resolution of the lytic lesion was evident on radiograph after 3 months of therapy. Clinical examination and radiograph after 2 years of therapy revealed natural shedding of deciduous molars and successful eruption of right mandibular premolars (fig 7) with complete resolution of the lesion (fig 8) and no surgical intervention was done.

DISCUSSION

The International Histiocyte Society in 1987 established a classification of histiocytosis in to three groups: (a) Langerhans cell histiocytosis (b) histiocytosis of mononuclear phagocytes other than Langerhans cells, (c) malignant histiocytic disorders. Even though this classification does not reflect recent neoplastic evidence, it is universally accepted. The vast majority of patients are developed

with the type a & b variety. The etiology and pathogenesis of LCH is still unclear. Two different hypotheses have been proposed:

- 1.) A disorder of immune regulation. The presence of aggregates of other immunologically active cells in lesions, a deficiency in the number of suppressor T lymphocytes and increased cytokines suggest an exuberant reaction of Langerhans cells to an unknown antigen or neoantigen⁵.
- 2.) A neoplastic process due to the monoclonal proliferation of Langerhans cells⁶. Other etiological factors have been suggested, such as viruses and trauma.

The stages in the condition have been classified as follows:

Stage I: A single bone lesion

Stage II: Multiple bone lesions (previously termed eosinophilic granuloma)

Stage III A: Bone + Soft tissue lesions often associated with diabetes incipidus and Exophthalmos previously termed Hand-Schuller-Christian triad

Stage III B: Soft tissue only (disseminated form)(previously termed Letterer Siwe disease)

These stages are not exclusive and some overlap or progression from one to the other can occur. The peak incidences of children from 1-15 years of age are the most commonly afflicted. The relative frequency of organ involvement is as follows: bone 80%, skin 60%, liver, lymph nodes 33% and maxillofacial 20%. Jaws are involved in less than 10% of children with the disease while mandibular involvement in young children is uncommon and bilateral affection is very rare. Studies have reported an incidence of 7.9% in the jaws with angle and body of the mandible.¹ The clinical course of LCH varies considerably depending on the extent and number of organ involved as well as the age of the patient at the time of diagnosis. Oral involvement is a frequent finding, but initial symptoms are non specific. Oral findings includes: gingival enlargement, oral ulceration, mobility of teeth with alveolar expansion, jaw pain, facial swelling as well as the classic intra-osseous lesions. Children with multiple organ and bone lesions commonly have mandibular involvement, alveolar bone loss and teeth that appear to be floating in air. Majority of the cases with mandibular involvement are over 20 years of age while in the present case, the child is 9 years old, which is a unique and rare occurrence. The diagnosis of LCH is based on morphological, immunohistochemical and ultrastructural features. Presence of Birbeck granules on electron microscopic examination is diagnostic. A most recent method for LCH diagnosis is the positive immunostaining for S-100 and the CD 1a antigen, both are positive in our presenting case. Definitive Management of LCH is very difficult due to wide heterogeneity of disease patients. Many therapeutic procedures are advocated to treat these lesions starting from conservative, non-surgical procedures like intralesional injections, radiotherapy, chemotherapy and also by surgical methods such as curettage, marginal resection. Most of studies

suggest that the management of LCH depends on prognostic indicator which includes: age, site involvement and organ dysfunctions. Age < 2 years, multiple sites involvement and organ dysfunction are suggestive of poor prognosis and difficult treatment management^{7,8}. Various evidences suggest that treatment of single system disease is generally benign and may include observation for spontaneous regression, radiation, surgical curettage or intralesional infiltration with steroids⁹. Evidence shows that chemotherapeutic agents which is known to be effective in patients with vital organ involvement or those not responding to more conservative therapy. Radiation therapy in children younger than 6 years of age has been associated with altered growth, arrested dental development as well as secondary tumour development. In this presenting case considering the age of child, a conservative treatment management was done with intralesional injections of steroids with complete resolution of the lesion without any surgical intervention.

CONCLUSION

A gold standard of management is careful clinical examination and good diagnostic skill which lead to earlier treatment with minimal deformity. Although unifocal lesions of LCH successfully respond to curettage, the possibility of recurrences or new lesions still remains which makes a long term follow up necessary. The purpose of this article is to present a case report of 9 year old child affected with LCH, which was managed conservatively and to enhance the understanding of LCH. This case report concludes that steroids injection therapy is a simple and minimally invasive procedure, with minimal risk of complications and has shown significant possible benefits in terms of improvement in pain, resolution of lesion; eruption of permanent teeth's and improves the quality of life of patient since no surgical intervention is done.

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Future Special Issues/ Symposia

Special Issues

- Injury/Trauma: An epidemic of Modern times

Symposia

- Influenza: A dangerous pandemic; needs to be defeated
- Parkinson's Disease