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Palatal Manifestation of Langerhans cell histiocytosis in a 20 month old child: A Rare presentation

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Abstract:

Background- Langerhans cell histiocytosis (previously known as histiocytosis X) is an inflammatory myeloid neoplasm commonly occurring in bones and skin but can involve other organs also, although it is a rare disease it is fairly common in young children. Most commonly seen in age group 1-3 years old.

Methods – A case of a 20-month-old boy with a chief complaint of growth in the hard palate since 3 months. On intraoral examination ulcerative growth was observed on hard palate extending from right side of 1st premolar to left side of 1st premolar. The ultrasonogram of whole abdomen was done which revealed cystitis. CECT scan of oral cavity was done and which showed ill-defined mildly enhanced soft tissue density mass.

Result – After serial examinations definite diagnosis of Langerhans cell histiocytosis was confirmed and surgical removal of bilateral lower part of the maxilla, teeth and hard palate with the primary lesion was done.

Conclusion-Despite all the major advances in our medical field it is difficult to understand and diagnose LCH. By doing detailed examinations it is possible to narrow down the diagnosis and planning accurate management.

Keywords: Palatal Manifestation

1. INTRODUCTION

Langerhans cell histiocytosis (previously known as histiocytosis X) is an inflammatory myeloid neoplasm commonly occurring in bones and skin but can involve other organs also ,although It is a rare disease it is fairly common in young children it is named so because the neoplastic cells resemble dendritic cells in skin[1] The incidence in children is approximately 5 cases per million and estimated 1 per million in adults.[2] Most commonly seen in age group 1-3 years old.[3]. According to SEER data in the United States there is a lower incidence in black people and higher in Hispanics [4,5] There is a presence of BRAF V600E mutation in more than half of cases, and key driver of this neoplastic disorder is MAPK (mitogen activated protein kinase. It can be seen as a multifocal or unifocal or can also present in multiple organ system and this differentiation is important in prognosis and treatment, however the cells being CD1a and CD207 + makes them different from their counterparts in the skin.[1] As per the classification given by WHO[2022] LCH is in the dendritic cell neoplasm group.[6] Diagnosis of this disease is done on the basis of histopathology. First manifestation of LCH Is oral Involvement

but mostly the symptoms are non-specific which can lead to misdiagnosis. The purpose of reporting this case is to discuss clinical, radiological, pathological features and the role of an ENT surgeon in diagnosing and management of this disease.[7]

2. CASE REPORT

A 20-month-old boy brought by his father presented to the department of ENT, tertiary care hospital with the chief complaint of growth on the hard palate for 3 months which was insidious in onset, gradually progressive and associated with difficulty in swallowing. Parents also observed weight loss which was not documented. There was a medical history of osteomyelitis of femur 1month ago which was treated at tertiary health center. No relevant history of trauma was given by the parent.

On general examination patient didn't have any pallor ,icterus ,cyanosis ,lymphadenopathy or clubbing Patient was conscious, B/L pupil reactive to light, recognizing parents, patient was irritable , vitally stable [RR -17/min HR-112bpm Sp02-98% on room air] On Physical examination we noticed scaling present on scalp of patient for which a dermatological referral was taken and treatment of taenia capitis was started. On intraoral examination a ulcerative growth, foul smelling, pinkish swelling of 3x3 cm was present on hard palate extending from right side of 1st premolar to left side of 1st premolar no bleeding or pus was seen from the region . On Palpation inspectory findings were confirmed. The swelling was non-tender, of normal temperature, and soft with diffused margins. No findings were detected in ear, nose and neck examination.

Investigations-Laboratory Investigations revealed WBC -10.5 X10^9/L, Hb -11.4gm/dl Plt-221x10^9/l. Liver Function Tests and renal function test were within normal limits. The ultrasonogram of whole abdomen was done which revealed cystitis. Pediatric reference was taken, and appropriate antibiotics were started. Chest examination was normal.

CECT scan of oral cavity was done and which showed ill-defined mildly enhanced soft tissue density mass measuring 30mmx32mm involving both half of hard palate and destruction of bony hard palate extending to left half of nasal cavity causing destruction of left upper alveolus was reported suggesting an overall neoplastic etiology .Contrast Enhanced MRI was also done and well defined lobulated heterogenous signal intensity mass lesion measuring 42 X 26 X 29 in midline ,hard palate infiltrating into floor of both nasal cavity extending into anterior premaxillary space showing moderate enhancement on contrast-s/o mitotic pathology with no extension into intracranial cavity. Biopsy was taken from the palate which revealed Langerhans cells histiocytosis with CD1a, S100 and CD 207 positivity in IHC which led to a final diagnosis of Langerhans cell histiocytosis of palate.

Management – Oral hygiene maintenance was advised**,** Benzocaine gel was prescribed for local applications 15min prior to each meal. Ryles tube was inserted and feeding of child was started. Preanesthetic clearance was taken from the department of anesthesia and separate clearance was taken from the department of pediatrics. Opinion was also taken in view of the post operative prosthetic

application from the department of prosthodontics Patient was posted for wide local excision of hard palate and bilateral infrastructure maxillectomy under general anesthesia. He was started on IV fluids and antibiotics and was kept nil by mouth 12hrs prior to surgery. General anesthesia was induced. Wide local resection of lesion with bilateral inferior maxillectomy with safe margins was done which were identified over the soft palate and in the upper gingivobuccal sulcus, superiorly maxillary sinus explored, and mucosa of maxillary sent for histopathological examination of margins and hemostasis achieved.The palatal defect was kept open to be healed by secondary intention and a merocele pack was placed in the left nasal cavity, to prevent nasal bleeding. GA reversal was uneventful.

Postoperative evaluation was done- patient was febrile (100degree F), RR-22/min HR-130bpm Sp02-97 percent on room air. Triple antibiotics with anerobic coverage and IV fluids started. Ryles tube feeding was started on postoperative day 2. Follow up after 15 days was done which revealed healthy granulation tissue and a clean wound margin.

3. DISCUSSION

A Multidisciplinary approach is required for careful examination and evaluation of Langerhans cell histiocytosis (LCH). In this condition there is a marked monoclonal proliferation of myeloid cells particularly histiocytes (Langerhans cells) followed by various plasma cells, eosinophils, lymphocytes and multinucleated giant cells.[8]. Mostly these cells are found in mucosa, lymph nodes, epidermis and bone marrow. According to literature it results in the destruction of hard and soft tissues due to monoclonal proliferation[9]. The etiology of disease is not clear yet and so it is difficult to treat this condition.

A wide age group is affected with LCH but children below age of 15 are more commonly involved with a peak occurrence at 2-4 years and it has twice male predilection as that of females[10].The present case is about a 20month old boy with disease involving the hard palate which was diagnosed on histopathology and IHC and was excised along with nferior maxillectomy .Oral manifestations are the first sign and symptoms in about 5-75% cases[11].Intraoral mass ,mucosal ulcers are the most common oral manifestations of LCH.[12] Differential diagnosis of LCH are Rhabdomyosarcoma, Langerhans cell sarcoma, Erdheim-Chester disease, Intermediate cell histiocytosis, Juvenile Xanthogranuloma, Multiple Myeloma, Hemophagocytic Lymphohistiocytosis and Rosai Dorfman disease.

Gardener et al suggested in his study that the patients with multifocal LCH or unifocal disease in CNS , prednisolone is the first line therapy for 1year, with the addition of mercaptopurine in high risk LCH [13]. In our case oral prednisolone (1-2mg/kg/day in 3 divided doses) was given for 1month. High risk LCH arises from the somatic mutation of hematopoietic progenitor cells while the low risk LCH arises from somatic mutation of tissue restricted dendritic cells .[14]

Krooks et al suggested in the study, the involvement of nails is less common but can be visible subungual pustules, purpuric striae, purulent discharge, hemorrhage, pitting .[15] The final diagnosis of

LCH is done based on histology and IHC where CD207, CD1a and S100 protein positivity is commonly seen.[16] In the present case, immunostaining was positive for CD1a, CD207 and S-100 which brought us to the definitive diagnosis.

Full body positron emission tomography is recommended in patients >1yrs old at baseline [1].The imaging modality of choice to visualize CNS involvement is Magnetic resonance imaging (MRI) with gadolinium contrast of brain in children which shows mass lesions or enhancement of pons, meninges, white matter of cerebellum, basal ganglia.[17]In another study 163 patients were reported out of them 29% were diagnosed with meningeal lesions and involvement of choroid plexus in 6%.[18] In the present case there was not intracranial involvement on MRI with gadolinium.

The treatment of LCH depends on the focality and the severity of the disease. Management includes a combination of surgical excision, chemotherapy and/or radiation therapy. In cases of single bone lesion, more commonly curettage is preferred, Radiotherapy (RT) is rarely administered to children. There are no prospective studies which have compared radiation therapy vs curettage. Systemic therapy should be started in cases where single bone lesions of more than 5cm or involvement of femur, vertebrae or CNS risk bones like mastoid, temporal, orbit, sphenoid bone.

In present case the disease was pertinent to maxillary bone the excisional curettage of the lesion was done taking maximum margins around the lesion, no bone grafting was done rather we opted for prosthetic management. Generally, in children chemotherapy is opted instead of radiotherapy with single agents like cytarabine or cladribine as RT is unsafe for osseous sites.[18,19]

4. CONCLUSION

LCH is a rare pathology, it is at high risk of being misdiagnosed .There are wide range of clinical symptoms in this disease so a thorough clinical history, examination with appropriate investigations ,thorough knowledge and coordinated multidisciplinary approach is essential in successfully managing the disease and to make a definite diagnosis at an early stage which will help in reducing morbidity and mortality associated with this condition.

CECT ORAL CAVITY- Axial image showing lobular shaped soft tissue density mass lesion involving hard palate causing erosion of surrounding bone. B- CECT ORAL CAVITY- Sagittal image showing heterogeneously enhancing mass lesion is seen involving hard palate.

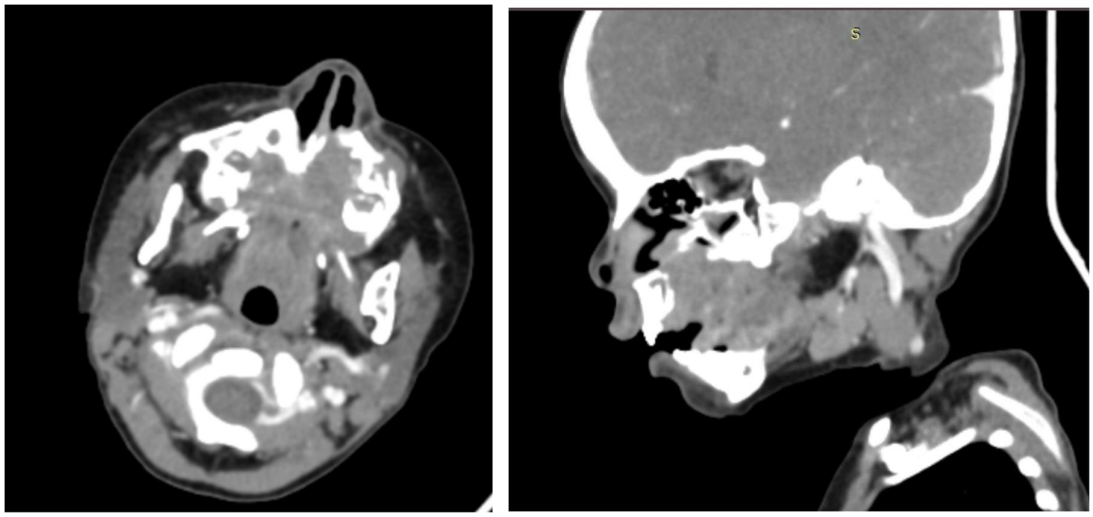
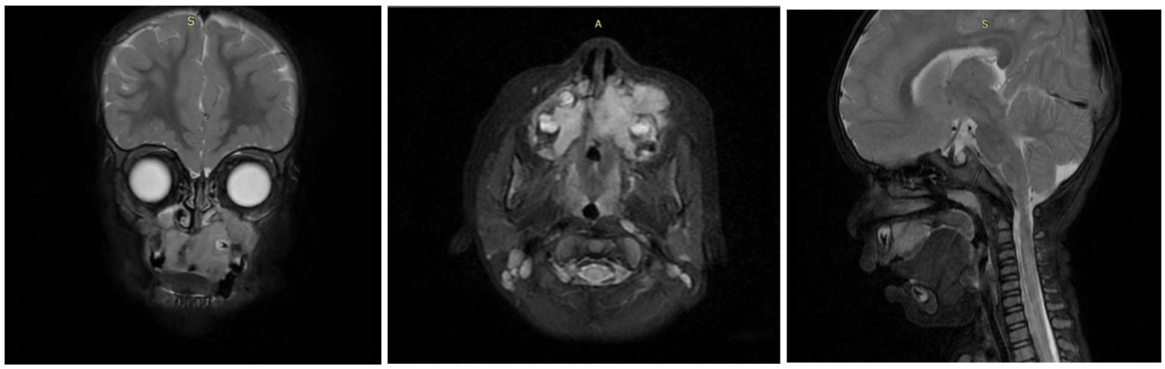


Figure . CEMRI ORAL CAVITY- T2W images (Axial, Sagittal and Coronal) -

Images showing irregular shaped hyperintense mass in the region of hard palate extending into nasal cavity. (Yellow arrow- hard palate mass, white arrow- normal soft palate.)



Intra- operative picture showing the involved excised hard palate with teeth, nasal septum and anterior nasal pack (merocele) placed for hemostasis.

Despite all the major advances in our medical field it is difficult to understand and diagnose LCH. By doing detailed examinations it is possible to narrow down the diagnosis and planning accurate management.

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