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Case Report

Palatal manifestations of Langerhan cell histicytosis in pediatric age: a case report with review of literature

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ABSTRACT

Langerhans cell histiocytosis (LCH) is characterised by multisystem disorder with various entities grouped together under same name. Oral cavity manifestations of this disease is yet to be explored. In oral cavity it can present with ulceration, swelling or osteolysis of underlying bone. Oral cavity involvement due to LCH might confuse the examining doctor resulting in misdiagnosis of such dreadful disease. So here we are reporting a case of LCH in a 4 years old female presented to us with ulceration over hard palate mucosa. Biopsy was taken from the ulcer site which was positive for LCH. On further examination and evaluation of the patient multisystem involvement was found with involvement of skull bone.

Keywords: Langerhan cell histiocytosis, Pediatric LCH, Palatal ulcer

INTRODUCTION

Langerhans cells are named after Paul Langerhans.[1] Various disease are grouped under Langerhans cell histiocytosis (LCH) namely histiocytosis X, eosinophilic granuloma, letterer-Siwe disease, Hand-Schuller-Christian syndrome, Hashimoto-Pritzker syndrome, self-healing histiocytosis, pure cutaneous histiocytosis, langerhans cell granulomatosis.[2] LCH is characterized by clonal expansion of myeloid precursors that differentiate into $CD1a^+/CD207^+$ cells in lesions leading to spectrum of organ involvement and dysfunction.[3] In the oral cavity LCH can present with mucosal ulceration and extensive osteolysis.[4] Here we report a case of LCH in a 4 years old female presenting with complaints of oral mucosal ulceration.

CASE REPORT

A 4 years old female presented to our outpatient department (OPD) with complaints of difficulty chewing food for last 1 year. Patient was also complaining of difficulty in walking for one and half years. There was past history of left sided ear discharge 1 year back which subsided on topical ear drops. On examination of face and head multiple lesions were present over the scalp along with single swelling over vertex of head. It was nearly 4×3 cm. No sinus or fistula over the swelling was seen. No redness over the skin was present. No visible pulsations were seen. On palpation inspectory findings were confirmed. Area was non tender with normal temperature. A soft swelling with diffused margins palpated. It was immobile and fluctuant (Figure 1). On examination of the oral cavity around 2×2 cm ulceration was seen over hard palate opposite 1st and 2nd molar which was indurated (Figure 2). Ear examination revealed bilaterally grade 1 retraction in the pars tensa. The patient was investigated further and her X ray skull was done which was showing a well-defined lytic punched out lesion caused by asymmetric destruction of inner and outer cortices resulting in characteristic bevelled edge. No periosteal reaction was seen (Figure 3). As patient also had pain over

left hip her x ray pelvis was done which was suggestive of subtle irregularity over left side acetabular cup (Figure 4).

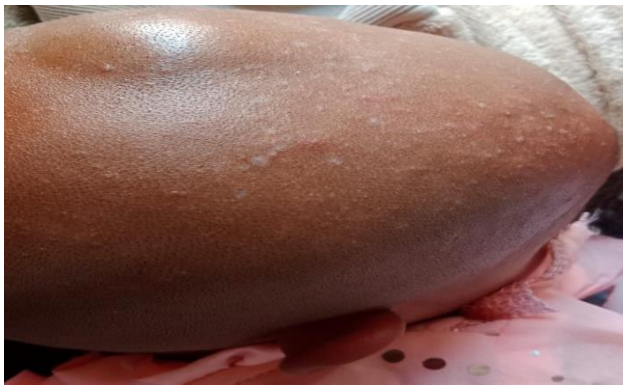

Figure 1: Showing swelling over the vertex along with multiple lesions over scalp.

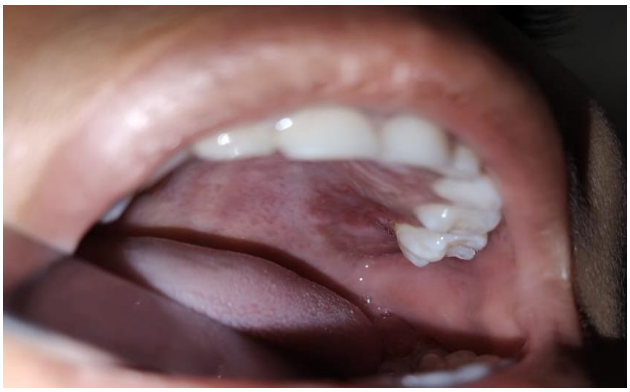

Figure 2: Showing ulceration over left hard palate.

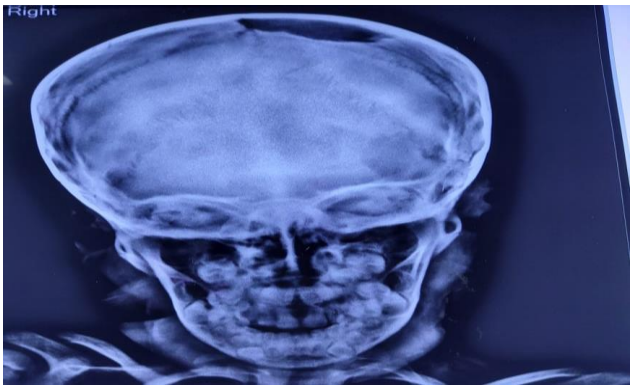

Figure 3: X ray skull showing well defined lytic punched out lesion.

Contrast-enhanced magnetic resonance imaging (CE-MRI) brain was showing evidence of well-defined oblong shaped lesions involving occipital bone on left side, mastoid part of left temporal bone, left parietal bone exhibiting iso to hypointense signal on T1W images and iso to hyperintense on T2W FLAIR images. On post contrast images significant enhancement of all lesions were seen (Figure 5).

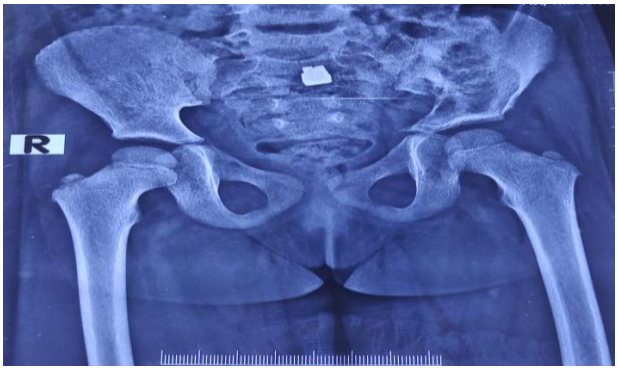

Figure 4: X ray pelvis was done suggestive of subtle irregularity over left side acetabular cup.

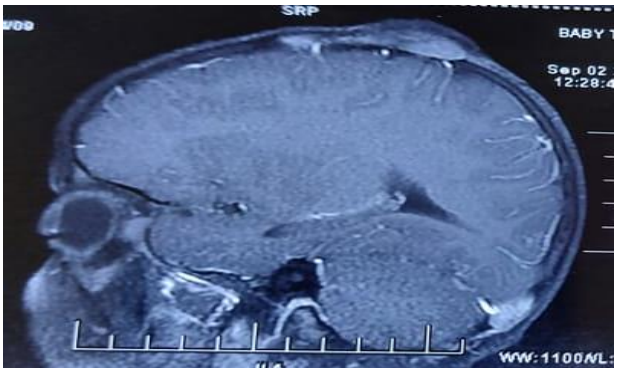

Figure 5: CEMRI brain was showing evidence of well-defined oblong shaped lesions.

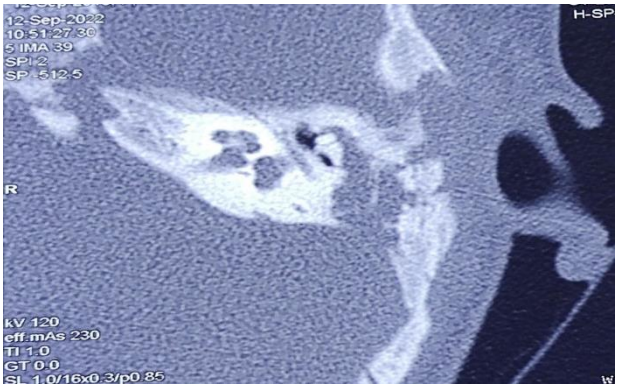

Figure 6: HRCT temporal bone was also advised for the patients which revealed multiple ill-defined osteolytic lesions.

High-resolution computed tomography (HRCT) temporal bone was also advised for the patients which revealed multiple ill-defined osteolytic lesions with few showing bevelled edges in left frontoparietal bone, occipital bone with squamous and mastoid part of temporal bone causing erosions of left mastoid air cells, posterosuperior wall of external auditory canal, scutum, sinodural plate, part of tegmen tympani and tegmen mastoideum with heterogeneously hyperdense soft tissue noted in skin an subcutaneous plane of left pinna and mastoid region and soft tissue contents in the left middle ear cavity, mastoid

antrum and residual mastoid air cells. Right ear revealed normal study (Figure 6). Biopsy from her oral cavity ulcer was taken which revealed subepithelium with plenty of Langerhans cells having abundant eosinophilic cytoplasm, elongated nucleus with nuclear grooves, fine chromatin and indistinct nucleoli in these cells. Many langerhan cells were multinucleated, occasional mitotic figures were also identified (Figure 7). On immunohistochemistry it showed positivity for CD1a. S-100 and negative for LCA. Features were suggestive of LCH (Figure 8). After the final diagnosis patient was referred to chemotherapy unit for further treatment. However, patient was advised multivitamin syrup along with maintenance of local hygiene. She was also prescribed benzocain gel local application 15 minutes before each meal for local anesthesia before chewing and swallowing food. Patient was followed up after 3 weeks and there was complete resolution of oral ulceration.

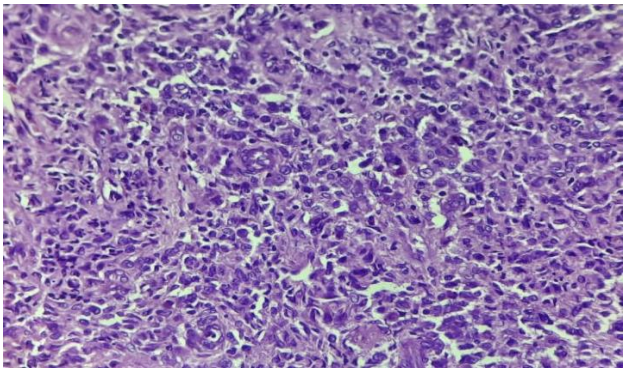

Figure 7: Haematoxylin eosin stain showing plenty of Langerhans cells having abundant eosinophilic cytoplasm.

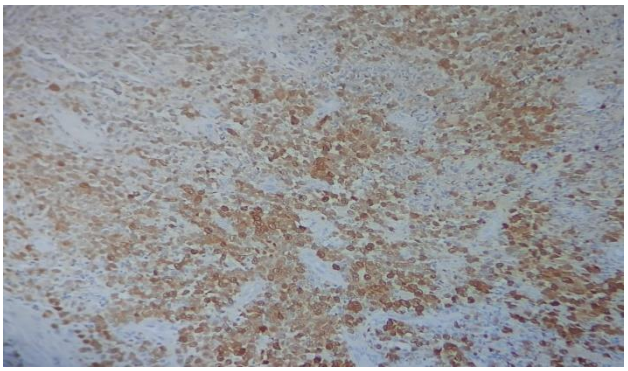

Figure 8: Immunohistochemistry showing positivity for S-100 protein.

DISCUSSION

LCH is a multisystem disorder requiring careful examination and evaluation from multidisciplinary department. Etiology of this disease is still not known which makes it difficult to treat this condition. According to Gadner et al current standard of care for front line therapy of patients with multifocal LCH or unifocal disease in CNS risk sites is vinblastine/prednisolone for 1 year, with the potential addition of mercaptopurine for high risk LCH.[5] Howarth et al in their study concluded that LCH generally follows the benign course. In patients in whom multisystem involvement has been confirmed or is suspected should be followed clinically after treatment for a minimum of 1 year but ideally all patients should be followed up for whole life to ensure continued remission. They also said that LCH is not specific for children, it should also be considered as differentials in case of adults with multisystem disease.[2]

Milián et al reported two cases of adult LCH limited to oral mucosa. In which one patient was first treated with perilesional triamcinolone acetonide infiltration (25 mg: 1 injection every 3 weeks for a total of 8 sessions) resulting to complete resolution of lesions. Lesions reappeared over right mandibular alveolar ridge and palate which also resolved with intralesional steroid. No new lesion appeared further on 1 year follow up. While in second case oral ulceration was treated via radiotherapy and after 3 year follow up no recurrence was seen.[6]

Merglova et al in their study described two case reports of oral cavity LCH in children's. In first case child had multifocal LCH and hence treated for same but after 12months of treatment recurrence was found for which child was treated using chemotherapy followed by radiotherapy and corticosteroid treatment while other child was treated according to procedure of skin and mucosa affecting form of LCH in paediatric oncology department.[7] The treatment of LCH is based on chemotherapy, radiotherapy and the administration of corticosteroids.[8] Isolated bony lesions are separately removed surgically.[9]

Our patient has multisystem involvement along with oral mucosal ulceration which was treated with conservative medical management. However, patient was referred for chemotherapy and further management in chemotherapy unit. On reviewing the literature, we found other reported cases of LCH with involvement of palate which is described in Table 1.

Table 1: Literature review of palatal LCH in children under the age of 4 years.[19]

Case	Author	Age	Sex	Lesion	Size (mm)	Ulcer	Type	Treatment	Prognosis
1	Roper et al[10]	9M	Undesc-ribed	Hard palate	25 (in diameter)	+	SS-s	Total excision	1Y FOD

Continued.

Case	Author	Age	Sex	Lesion	Size (mm)	Ulcer	Type	Treatment	Prognosis
2	Loh et al[11]	2Y	Male	Hard palate + upper gingiva	4 (in diameter)	-	MS (skin of scalp)	Total excision +chemotherapy CRT at recurrent	4Y FOD
3	Davis et al[12]	9M	Female	Hard palate + upper gingiva	30×10 15×10	-	SS-m	Chemotherapy	Developing normally
4	Alajbeg et al[13]	2.5 Y	Male	Palate + upper gingiva	Not described	+	MS (skin + cranium)	Chemotherapy	Not specified
5	Sasaoka et al[14]	10 M	Female	Upper gingiva	8 (in diameter)	-	MS (skin and cranium)	Chemotherapy	2.5Y FOD
6	Murray et al[15]	22 M	Male	Hard palate + upper gingiva	20×5	+	MS (cranium and mandible)	Chemotherapy	6 week FOD
7	Merglova et al[7]	13 M	Female	Upper gingiva	Not described	+	MS (skin of abdomen and scalp)	Chemotherapy CRT at recurrent	8Y FOD
8	Merglova et al[7]	5M	Undesc-ribed	Upper gingiva	Not described	+	MS (skin of abdomen)	Sent to another hospital	Undescri-bed
9	Halperson et al[16]	2Y	Male	Upper gingiva	12,15 (in diameter)	+	MS	Chemotherapy	1Y FOD
10	Nevessilva et al[17]	2Y	Male	Hard palate + upper gingiva	Not described	-	Undescri-bed	Sent to another hospital	Undescri-bed
11	Nevessilva et al[17]	2Y	Male	Hard palate + upper gingiva	Not described	+	Undescri-bed	Sent to another hospital	Undescri-bed
12	Hammouri et al[18]	2 Y	Male	Upper gingiva	Not described	+	Undescri-bed	Sent for chemotherapy	Undescri-bed
13	Togawa et al[19]	4M	Male	Upper gingiva	10 (in diameter)	-	SS-s	Total excision	2Y FOD
14	Present study	4 Y	Female	Hard palate	20 x 20	+	MS (hard palate + cranium)	Sent for chemotherapy	On follow up

SS-s: single system, single site, SS-m: single system, multisite, MS: multisystem, FOD: free of disease.

CONCLUSION

Oral manifestations of LCH is difficult to diagnose and may even get unidentified. Every ENT surgeon must be well aware of ENT manifestations of LCH so that if any suspected patient comes thorough investigations can lead to early diagnosis of such patients.

It can also unfold other system that are involved leading to better patient care and thus reducing the morbidity.

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