

A Rare Case Presentation on Langerhan's Cell Histiocytosis an Chronic Disseminated Form with Gingival Enlargement in Three and a Half Year Old Pediatric Patient

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Abstract

Langerhan's Cell Histiocytosis formerly known as histiocytosis X traditionally denotes a group of diseases that stem from proliferative reticuloendothelial disturbances. The etiology and pathogenesis of the disease remain debatable. The present paper reports a case occurring in a three and a half year old (3½ year pediatric) child reported to the department of oral pathology with gingival enlargement of the jaws, also discussed are the radiological features along with histopathological features of the case.

Keywords: Histiocytosis X; Langerhan's cell; Histiocytosis; Exophthalmos; Diabetes insipidus

Introduction

Langerhan's cell histiocytosis (LCH), formerly known as Histiocytosis X is currently considered as disorder of immune regulation manifested by abnormal proliferation of histiocytosis and granuloma formation.

The Langerhan's cell, a unique histiocyte is the distinctive pathologic component of the disease. LCH may affect any organ, although the reticuloendothelial system (i.e. bones, skin, lymph nodes, liver and spleen) is involved in most cases [1].

Langerhan's cell disease manifests in three forms:

Acute disseminated form: It is previously referred to Letterer-siwe-disease most likely represents a malignant neoplastic process. It is characterized by a rapidly progressive, clinical course and widespread in organs, bones and skin involvement by the proliferative process in infants has been the common presentation.

Chronic disseminated form: It is previously referred to as Hand-Schullers-Christian syndrome with the classical triad of lytic bone lesions, exophthalmos and diabetes insipidus.

Chronic Localized form: With only unifocal or multifocal bone lesions previously termed as eosinophilic granuloma [2].

Etiopathogenesis: The pathogenesis of LCH is unknown. It may be caused due to hypersensitive reaction to unknown antigen with stimulation of histiocytes-macrophage system.

Deficiency of suppressor lymphocytes (T-8, altered immunoglobulin's autoantibodies and structural changes to thymus in all the advanced forms have been found in LCH patients.

An inflammatory origin is also suspected due to the microscopic characteristics and clinical evolution, a bacterial origin although no specific causal microorganism has been identified [3].

Here I report a case of chronic disseminated form of Langerhan's cell disease (Hand-Schullers-Christian disease) presenting with gingival enlargement in 3½ year old pediatric patient.

Case History

A three and half year old male child was referred to department of oral pathology with swelling, pain and mobility involving deciduous canine and first molar region involving all the four quadrants since five months. The patient's father further gave the history of ulcers and inability to tolerate hot food. There was no history of trauma or any associated family history.

General examination of the patient with paediatrician and ENT surgeon revealed presence of Hepatosplenomegaly and Otitis media (Figure 1).



Figure 1: Preauricular pus discharge suggestive of Otitis Media.

Extra oral examination revealed right submandibular lymphadenopathy however there was no exophthalmos. Intraorally gingival enlargement was seen in molar region in all the four quadrants. Each enlargement was around 1.5 2 cm in size, diffuse

reddish in colour and soft to firm in consistency, bleeding was evident on slightest provocation (Figure 2).



Figure 2: Intra oral pictures showing gingival enlargement.

Radiographic examination showed punched out lytic lesions in the skull involving parietal, temporal and frontal bones. In jaws there was diffuse destruction of bone and displacement of teeth (Figure 3).



Figure 3: Radiograph (Lateral view) shows multiple punched out lesions in skull.

Lab investigations were within normal limits which included CBC, LFT, coagulation studies and urine osmolality.

Microscopic examination of the lesion revealed presence of many histiocyte like cells, eosinophils and lymphocytes, diffusely scattered throughout the connective tissue stroma, keeping the clinical, radiological and histopathological features in mind a diagnosis of Langerhan's cell disease (Histiocytosis X) in general and Hand-Schullers-Christian disease in particular was made (Figures 4 and 5).

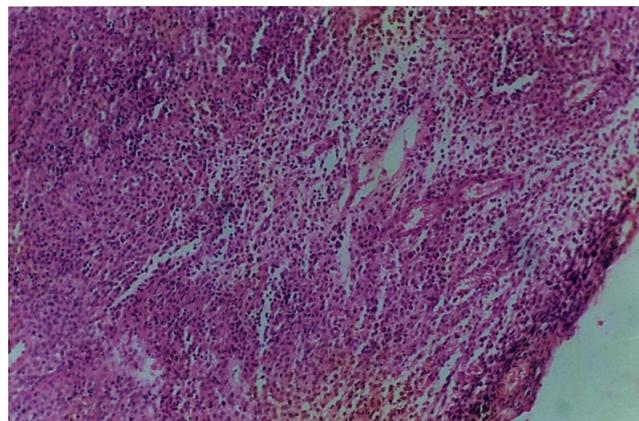


Figure 4: H and E section shows numerous histiocyte like cells, eosinophils and lymphocytes in loose connective tissue stroma (low power; 10X).

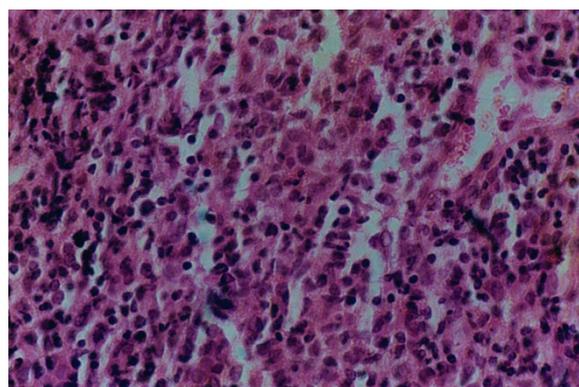


Figure 5: H and E section shows numerous histiocyte like cells, eosinophils and lymphocytes in loose connective tissue stroma (high power; 40X).

The patient was kept on prednisolone 5 mg q.i.d for three weeks with supportive treatment consisting of antibiotics and eardrops.

On subsequent follow up patient had ear discharge and swelling in head region and was advised bone biopsy. Patients bone biopsy suggested Histiocytosis X and patient was advised regular follow up for periodic review. But the patient did not come for subsequent follow up.

Discussion

Langerhan's cell histiocytosis a chronic disseminated forms i.e. Hand-Schuller-Christian disease is characterized by widespread skeletal and extra skeletal lesions and a chronic clinical course. It occurs usually before the age of five but has been reported even in young adults. It's more common in boys with a gender ratio approximately 2:1. In Hand-Schullers-Christian disease the skeletal system and soft tissues may be involved while in eosinophilic granuloma only the bone is affected. Although soft tissue extension is often observed. Letterer-Siwe disease is an acute fulminating disease with

widespread lesions of both skeletal and extra skeletal tissues including the skin.

Langerhan's cell histiocytosis and chronic disseminated form is characterized by a classic triad of single or multiple punched out bone lesions in the skull, unilateral or bilateral exophthalmos and diabetes insipidus with or without other manifestations like polyuria, dwarfism or infantilism. The complete triad is seen only in 25% of the affected patients. Involvement of facial bones is frequently associated with soft tissue swelling, tenderness and facial asymmetry. Otitis Media is also common. Other bones frequently involved are femur, ribs, vertebrae, and pelvis. Sometimes the skin exhibits papular or nodular lesions.

Oral manifestations are the earliest signs reported in around 5-75% of patients. These are often nonspecific and include sore mouth, halitosis, gingivitis, unpleasant taste, loose and sore teeth with their early exfoliation and failure of extracted tooth sockets to heal. Leading to loss of supporting bone mimicking advanced periodontal disease.

Radiographic examination reveals that the individual lesions particularly in the skull are sharply outlined and those in the jaws may be more diffuse exhibiting destruction of alveolar bone with tooth displacement.

Histologically Langerhan's cell histiocytosis a chronic disseminated form (Hand-Schullers-Christian disease) manifests in 4 states during its progression:

- A proliferative histiocytic phase with accumulation of collections of eosinophilic leukocytes scattered throughout the sheets of histiocytosis.
- A vascular granulomatous phase with persistence of histiocytes and eosinophils, sometimes with aggregation of lipid-laden (cholesterol), macrophages.
- A diffuse xanthomatous phase with abundance of "foam cells"
- A fibrous or healing phase [4].

Electron microscopic evaluation of the lesional tissue has been the gold-standard Ultrastructurally; Langerhan's cells contain rod shaped cytoplasmic structures known as birbeck granules which differentiate them from other mononuclear phagocytes. Langerhan's cells show immuno reactivity to CD-1a or CD-207, the latter marker being even more specific to Langerhan's cells. In few cases lesional cells have shown immunoreactivity to S-100 protein and peanut agglutinin (PNA)[5].

Lab investigations often reveal anaemia, leukopenia and thrombocytopenia. The serum cholesterol level is nearly normal. Although tissue cholesterol content may be elevated remarkably.

Treatment of choice is curettage or excision of lesions, inaccessible lesions may be irradiated. Some patients are benefited from chemotherapeutic drugs including prednisolone. One of the significant factors influencing the morbidity and mortality of the disease is the extent of the disease at the time of initial diagnosis and number of organs systems involved [4].

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