Review Article

Langerhan Cell Disease of the Jaw: A Review

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Received: 01-09-2013

Revised: 01-10-2013

Accepted: 12-10-2013

Abstract:
Langerhan cell disease(LCD) is a disorder characterized by a proliferation of cells exhibiting phenotypic characteristics of langerhan cells. The natural history of the disease varies from a slow, benign, localized symptomatic bony or soft tissue lesion, to a rapidly progressive widespread multiple organ disorder which is often fatal. It is usually seen in the paediatric age groups and rarely seen in adults. A very few studies have focused on the clinical features and treatment in adult patients. It has been stated that isolated LCD of the jaws is associated with multiorgan disease and multifocal jaw involvement is associated with focal lesions in another organs. This review focuses on the clinical and histologic features of langerhan cell histiocytosis and also on the correlation of unifocal and multifocal jaw involvement with that of other organs.
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Keywords: Langerhan cell disease, Langerhan cells, Multiorgan disease, South East Asia

This article may be cited as: C Sreeja, P Gobu, Aesha, Tanveer S. Langerhans Cell Disease of The Jaw : A Review. J Adv Med Dent Scie 2013;1(2):97-100.

Introduction:

Langerhan cell disease (LCD) is a rare proliferative disorder that results in accumulation of pathologic langerhan cells in one or multiple organs or tissues[1,2,3]. It can present at any period ranging from birth to old age with a peak between 1-3 years. The incidence in the paediatric range has been estimated at 3-4 per million and 1-2 adult/million population with an overall incidence of two to five cases per million yearly[,4,5]. It has been stated that isolated LCD of the jaws is associated with multiorgan disease and vice versa[6].

This review focuses on the clinical and histologic features of langerhan cell histiocytosis and also on the correlation of unifocal and multifocal jaw involvement with that of other organs.

Discussion:

Review on the English literature gives an idea that it is a disease of younger adult,[7,8] and gender ratio is equal in all LCD cases . Studies report that males outnumber females (ratio of 1.5:0.8[6]1.8:1[9]).

Recently a prospective study from Rome report that multisystemic organ involvement is associated with a unifocal involvement in the jaws and multifocal involvement in the jaw is associated with a unisystemic involvement of the disease.[6] Table1 depicts the various reports of adult cases of isolated lesions involving Facial bones with no systemic involvement reported around the globe. Details obtained on an archival search.

On a review of literature, pain and mobility is the chief presenting complaint of patients with eosinophilic granuloma of bone. Other clinical symptoms include mobile teeth within the affected areas most commonly the posterior teeth, tooth ache headache and sensory disturbances.[9,5,10,11,12]

Few studies report that the Langerhan cell histiocytosis appear as an osteolytic lesion with diffuse and ill defined borders and in most of the reported lesion the borders are well demarcated with " floating teeth appearance.[3,8]

The most common differential diagnosis given for the characteristic radiographic appearance are cyclic neutropenia, large periapical cyst , odontogenic cyst and tumors, osteomyelitis, multiple myeloma, giant cell granuloma, aggressive periodontitis, deep fungal infection, necrotizing sialometaplasia and gaint cell granuloma.[1,11,14]

The histopathologic features of LCD are well characterized and recognized readily by oral and maxillofacial pathologist. The typical lesion is composed of an admixture of Langerhan cell histiocytes and interdigitating cells of dentritic cell lineage, T- cell lymphocytes, eosinophils and macrophages. Occasionally giant cells can be seen.[15,16]

CD1a is the specific marker for normal Langerhan cells and pathologic cells in LCD. The monoclonal antibody reactive to CD1a is is effective for immunohistochemical analysis of formalin- fixed tissue, replacing the less specific anti S-100 protein for the confirmation of LCD as S-100 shows positive reaction to melanocytes, langerhan cells and neural cells, .

Other markers that show positive rections to lagerhan cells are vimentin, langerin(CD207), Fascin, CD74, Peanut agglutinin lectin macrophage associated antigen,CD68 and Catepsin E[1,17].

There are several accepted treatment for langerhan cell disease. Surgical curettage, local irradiation and chemotheraphy (methotrexate and vinblastine with prednisone) have been used alone or in combinations. Some studies have reported good response to intraosseous steroid injections.[11,8,13]

Conclusion:

The various the clinical and histologic features of langerhan cell histiocytosis and also on the correlation of unifocal and multifocal jaw involvement with that of other organs has been discussed. This correlation helps in better diagnosis and treatment of the patient but further review is required to confirm the same.

Table1: Adult cases of isolated lesions involving Facial bones with no systemic involvement reported around the globe.

Age (In Years)	Gender	Year of report	Site	Author	Place of report
Above 18	Not specified	1997	N=14 Maxilla=8 Mandible=6	Iris Baumgartner et al[16*]	Switzerland
45	Female	2004	Mandible	Steven J.et al[18]	London (UK)
57	Male	2006	Skulll and orbit	Panagiotis V Kitsoulis et al[7]	Greece
16	Male	2007	Orbital region	José Antonio García et al[15]	Cordoba. Spain
28	Male	2007	Right upper maxilla (from 12 to 17).	José Antonio et al[15]	Cordoba. Spain
17	Female	2002	Frontal sinus	Basha et al[19]	Northern Ireland, U.K.
52	Female	2001	Ulceration lingual to the mandibular left first molar, no jaw involvement in radiograph	Milián MA et al[11]	London
Not specified	Not specified	2009	N=7 (maxilla and mandible	Annibali S et al[6]	Rome, Italy
28	Female	1997	Right mandibular mass	Fiorina Giona et al[20]	Rome, Italy
24	Female	1997	Right mandibular mass	Fiorina Giona et al[20]	Rome, Italy
38	Male	1997	Left mandibular mass	Fiorina Giona et al[20]	Rome, Italy
20	Male	1997	Left temporal bone mass	Fiorina Giona et al[20]	Rome, Italy
Above 13	N= 25; M = 17 F= 8	1999	Mandible= 26 Maxilla = 15	Ardekian et al[11]	Haifa,Israel

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Source of support: Nil

Conflict of interest: None declared