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Case Report

Cheilitis granulomatosa- A case report in an orthodontic patient

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ABSTRACT:
Cheilitis granulomatosa is a rare inflammatory disease with classical clinical features of diffuse, non-tender swelling involving one or both the lips. It might be a part of some syndromes. Arriving at a diagnosis is a challenge to the dentist. This case report attempts to throw some light on this rare disease.

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INTRODUCTION

Cheilitisgranulomatosa (CG) is an inflammatory disease with classical clinical features of diffuse, non-tender swelling involving one or both the lips. This rare disorder was first reported by Miescher in 1945.[1]CG is oftenassociated with Merkelson Rosenthal syndrome(MRS) to form a triad alongwith facial nerve palsy and fissured tongue (lingua plicata).[2]However, only rarely all the three symptoms of the triad are present together because of which the diagnosis is established after several years of follow up.[3]Both CG and MRS werecategorised as subsets of Oro FacialGranulomatosis (OFG) by Wiesenfeld in 1985.[4]Episode of recurrent lip swelling is gradually followed by a permanent enlargement of the lips along with changes in the oral mucosa. Though it can occur at any age, it is most commonly seen in the second and third decade of life with no gender predilection.[5]

The histological picture presents with non-necrotising granulomas, oedema, perivascular lymphatic infiltration and lymphangiectasia.[6] The mere histological findings are neither conclusive nor a prerequisite for the diagnosis. The diagnosis can usually be made by correlating the patient's history and the clinical features.[7] As the aetiology of CG is unknown, various treatment modalities have been proposed, however the effectiveness is limited, with the first line of treatment being intralesionalglucocorticoids.[4, 8] The differential diagnosis may include post traumatic swelling, neoplasm, actinic cheilitis, amyloidosis, angio-oedema, sarcoidosis or erythema multiforme.[9]

CASE REPORT

A 25 years old female patient reported with the chief complaint of swollen and thick upper and lower lips since the last 15 years, undergoing orthodontic treatment. No history of allergy, pain, facial paralysis, loss of taste sensation, burning sensation or dryness of mouth was reported. There was no significant past medical or familial history. She had not undergone any treatment for the swollen lips other than the orthodontic treatment reported. The swelling of the

lips was diffuse, soft and non-tender. Intra orally, generalised gingival hyperplasia was present along with a fissured tongue. Though the diagnosis of CG could be made on the basis of history and clinical presentation, punch biopsy was done to rule out other diseases like amyloidosis or neoplastic diseases. (Fig. 1- 3)

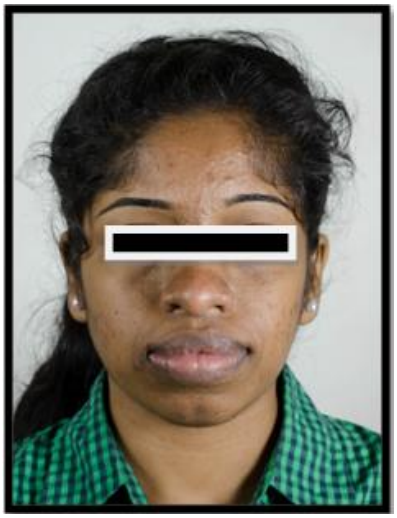

Fig.1- Frontal profile view

Fig. 2- Lateral profile view

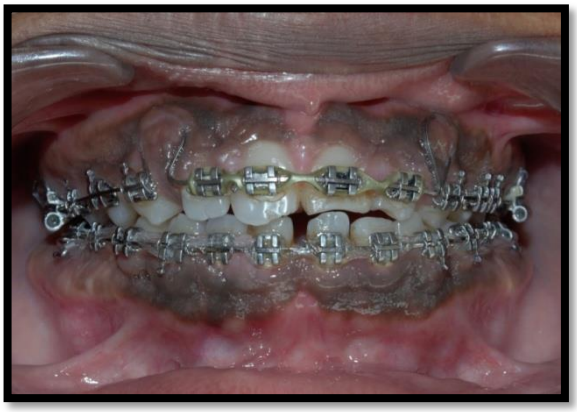

Fig. 3- Intraoral view

HISTOPATHOLOGY

The biopsy showed hyperplastic epithelium of variable thickness. The superficial lamina propria was oedematous and loose. The underlying fibrocellular connective tissue had plenty of non-caseating granulomas with Langhans type multinucleated giant cells and perivascular lymphocyte aggregations [Fig. 4].

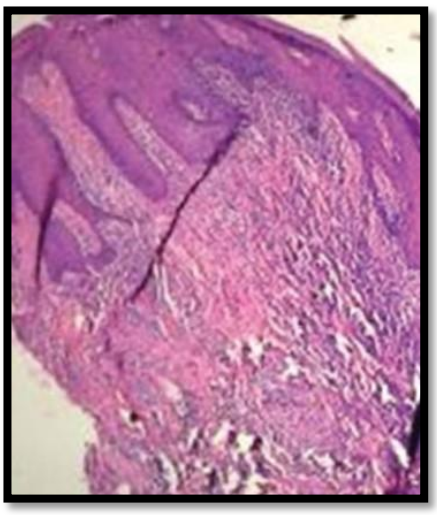

Fig.4- Histopathological view

Chest radiograph, Mantoux test, allergy and sensitivity tests did not show any significant result.

DISCUSSION

CG is a chronic, persistent, painless swelling of one or both the lips with distinct clinicopathologic features. Spontaneous resolution is seen very rarely. It is a chronic,non-necrotising granulomatous inflammation in the submucosal connective tissue.Mysterious etiology of CG includes an autosomaldominant inheritance pattern (responsible gene mapping to chromosome 9 p11)[10], allergic reactions[11], chronic infectious odontogenic foci, autoimmunemechanism, as an association with Crohn's disease and sarcoidosis or even as oral manifestation of systemic diseases .[12]Finally, the diagnosis of CG isachieved by correlating the patient's history and clinical features alongwith the histopathological findings.

Since the etiology is not known, effectiveness of treatment is limited. It is often misdiagnosed as an allergic reaction, infection, Crohn's disease or sarcoidosisleading to alternate treatments with limited results.[4] Crohn's disease and sarcoidosis are generally associated with systemic signs and symptoms making the diagnosis more definitive.[13] It is oftenassociated with Merkelson Rosenthal syndrome(MRS) alongside facial paralysis and fissured tongue. Individual presentation of CGwithout lingua plicata or facial palsyis described as "oligosymptomatic" or monosymptomatic form or an incompletevariant of Melkersson-Rosenthal syndrome orMiescher syndrome or Miescher'sCheilitis.[14, 15]Diagnostic tests

such as chest radiograph, AFB stainingand Mantoux test are required to exclude other etiologies of granulomatous disease, such as sarcoidosis or Mycobacteriuminfection. Gastrointestinal tract endoscopy and contrast radiography are helpful to exclude Crohn's disease. Patch tests/ allergy sensitivity test can rule out the allergens and the food additives in the pathogenesis of CG.

The first line of treatment includes topical, intra-lesional or oral corticosteroid therapy, however the relief is only temporary leading to recurrences.[16] Treatment with an anti-TNF agent (mainly infliximab) is the most recommended therapeutic option for granulomatous cheilitis (with or without CD) after failure of conventional treatments.[17, 18]. Photobiomodulation (PBM) using Low-Level Laser Therapies (LLLT) have proven to have significant anti-inflammatory properties and facilitate tissue repair.[19] Significant reduction of the lip oedema, with reduced transverse measurements and reduced thickness with values close to 35% have been achieved with PBM with no recurrence.[18]Cheilioplasty should be used as the last resort in severely disfiguring cases only after the disease has reached a quiescent phase followed by a support phase of intra-lesional therapy to prevent relapse.[20]

CONCLUSION

Since the diagnosis of CG isachieved by correlating the patient's history and clinical features alongwith the histopathological findings, the expertise of the clinician plays a great role in understanding the lesion and its treatment.

REFERENCES

1. Miescher G. ÜberessentiellegranulmatöseMakrocheilie (Cheilitisgranulomatosa). Dermatologica 1945; 91: 57–85.
2. Hornstein OP. Melkersson-Rosenthal syndrome: a neuromucocutaneous diseaseof complex origin. CurrProbl DermatoI1973;5:117-156.
3. Greene RM, Rogers RS, 3rd. Melkersson–Rosenthal syndrome: a review of 36patients. J Am AcadDermatol 1989; 21: 1263–1270.
4. Wiesenfeld D, Ferguson MM, Mitchell DN, MacDonald DG, Scully C, Cochran K, *et al*. Oro-facial granulomatosis: A clinical and pathological analysis. Q J Med 1985;54:101-113.
5. Zimmer WM, Rogers RS, Reeve CM, Sheridan PJ. Orofacial manifestations ofMelkersson–Rosenthal syndrome. A study of 42 patients and review of 220 casesfrom the literature. Oral Surg Oral Med Oral Pathol 1992; 74: 610–619.
6. Gojkov-Vukelic M, Hadzic S, Kantardzic A. Cheilitisgranulomatosa. Med Arh.2011;65(6):373-374. PMID: 22299304.
7. RIF Van Der Waal; EAJM Schulten; MR Van De Scheur; IMPMJ Wauters; TMStarink; I Van Der Waal (2001). Cheilitisgranulomatosa. , 15(6), 519–523.doi:10.1046/j.1468-3083.2001.00353.x
8. Shafer WG, Hine MK, Levy BM. A textbook of oral pathology. Philadelphia: WBSaunders, 1983:18.
9. Scully C, Langdon J, Evans J. Marathon of eponyms: 13 Melkersson-Rosenthal syndrome. Oral Dis 2010;16:707-708.
10. Muellegger RR, Weger W, Zoechling N, Kaddu S, Soyer HP, El Shabrawi-Caelen L, *et al.* Granulomatous cheilitis and Borreliaburgdorferi: Polymerase chain reaction and serologic studies in a retrospective case series of 12 patients. Arch Dermatol 2000;136:1502-1506.
11. Lazarov A, Kidron D, Tulchinsky Z, Minkow B. Contact orofacialgranulomatosis caused by delayedhypersensitivity to gold and mercury. J Am AcadDermatol2003;49:1117-1120.
12. Frankel DH, Mostofi RS, Lorincz AL. Oral Crohn's disease: report of two cases inbrothers with metallic dysgeusia and a review of the literature. J AmAcad Dermatol1985;12:260-268.
13. Worsaae N, Pindborg n. Granulomatous gingival manifestations ofMelkersson-Rosenthal syndrome. Oral Surg1980;49:131-138
14. Rintala A, Alhopuro S, Ritsila V, etaI. Cheilitisgranulomatosa. The Melkersson-Rosenthal syndrome. Scand J PlastReconstrSurg1973;7:130-136
15. Williams PM, Greenberg MS. Management of cheilitisgranulomatosa. Oral SurgOral Med Oral Pathol 1991; 72: 436–439
16. Krutchkoff D, James R. Cheilitisgranulomatosa: successful treatment with combined local triamcinolone injections and surgery. Arch Dermatol 1978; 114:1203–1206.
17. Barry O, Barry J, Langan S, Murphy M, Fitzgibbon J, Lyons JF. Treatment of granulomatous cheilitis withinfliximab. Arch Dermatol 2005;141:1080-1082.
18. Taxonera, Carlos; Alba, Cristina; Colmenares, Michel; Olivares, David; Rey, Enrique. Recurrent granulomatous cheilitis associated with Crohnâ€™s diseasesuccessfully treated with ustekinumab: case report and literature review. TherapeuticAdvances in Gastroenterology, 2020;13(), 175628482093432–.doi:10.1177/1756284820934327
19. Garagiola, U., Rossi, M. et al. Photobiomodulation and Miescher's cheilitisgranulomatosa: case report. MaxillofacPlastReconstrSurg2020;42: 35.https://doi.org/10.1186/s40902-020-00279-y
20. Kruse-Lösler B, Presser D, Metze D, Joos U. Surgical treatment of persistent macrocheilia in patients withMelkersson-Rosenthal syndrome and cheilitisgranulomatosa. Arch Dermatol 2005;141:1085-91.