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CASE STUDY

CHEILITIS GRANULOMATOSA - AN ENIGMA TO CLINICIANS

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ABSTRACT

Cheilitis granulomatosa is a chronic swelling of the lip due to granulomatous inflammation. It is a rare inflammatory disorder first described by Miescher in 1945. It is a monosymptomatic form or an incomplete variant of Melkersson-Rosenthal syndrome; a triad of recurrent orofacial edema, recurrent facial nerve palsy and fissuring of the tongue. As the etiology remains unknown, treatment of cheilitis granulomatosa is very challenging. We here by present a case of cheilitis granulomatosa in a 42-year-old female patient with swelling of lower lip for last eight years. This case highlights the importance of extensive investigations in the diagnosis of this lesion as the findings mimic many other granulomatous conditions.

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INTRODUCTION

Cheilitis granulomatosa is a rare and unique disorder. This condition is characterized by chronic swelling of one or both the lips due to granulomatous inflammation. It is a rare inflammatory disorder first described by Miescher in 1945. (Worsaae *et al*., 1982; van der Waal *et al*., 2001) It is a monosymptomatic form or an incomplete variant of Melkersson-Rosenthal syndrome (MRS); a triad of recurrent orofacial edema, recurrent facial nerve palsy and fissuring of the tongue. Cheilitis granulomatosa is also considered a subset of an uncommon disease Orofacial granulomatosis, which was introduced by Wiesenfeld in 1985. (Wiesenfeld *et al*., 1985) CG usually affects young adults, mostly in the 2nd decade of life with a female predilection. The estimated incidence of CG is 0.08incidence of the population.

Case History

A female patient aged 42 years reported to our clinic with chief complain of longstanding swelling of her lower lip (Figure 1) along with dental pain. On further examination soft, painless swelling was present in her lower lip. Patient complained that the swelling was present for last 8 years, for that she had gone through several medications but swelling did not disappear.

Further examination was carried out

ON INTRAORAL EXAMINATION Patient had multiple missing teeth, chronic periodontitis and multiple mobile teeth as well. Tongue was apparently normal .Her oral hygiene was extremely poor.

Then patient was advised to go for opg x-ray to evaluate the dental conditions. XRAY OPG (Fig. 3) Suggested multiple missing teeth, vertical and horizontal bone loss and all four last molars were found impacted

On extra oral examination

Diffuse swelling was noticed in the lower lip .Swelling was painless in nature. Soft in consistency, non tender there was no cracks or fissures were present. Colour of the lip was looking apparently normal. As patient told us there was never bleeding from lip swelling and not any other difficulty.

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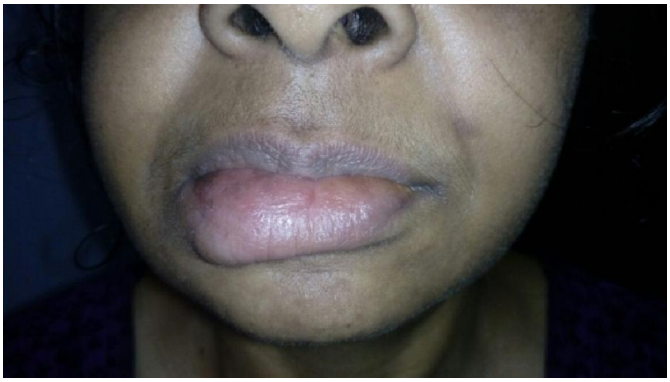

Fig. 1. Patients extraoral view

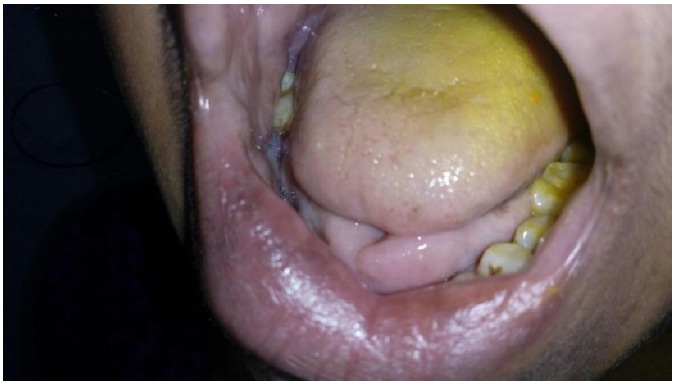

Fig. 2. Patient's intraoral view

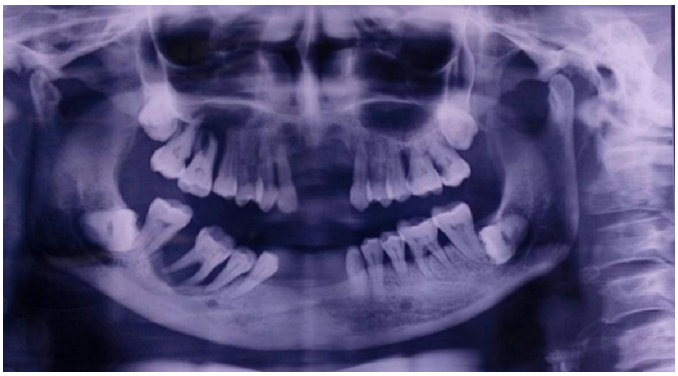

Figure 3. Patient's orthopantomograph

Patient's past history

Patient further told that swelling started 8 years back suddenly in the morning, she never noticed such type of swelling before that. Then she contacted local doctor for the treatment. She was being treated with antihistaminics drugs and steroids suspecting of allergic reactions. But swelling did not disappear even after 2 months of therapy. Then she contacted various specialists for the treatment but she did not get any relief. Lateron she was suggested to go for histopathology of the swelling. BIOPSY of lower lip was carried out. BIOPSY specimen suggested feature of skeletal muscles and fibrocollagenous tissues covered by acanthotic, focally parakeratotic stratified squamous epithelium with several discrete granulomas extending from subepitheilum to deeper structures. The granulomas were composed of plump histiocytes, lymphocytes, plasma cells along with multinucleated giant cells. There was no foci of casseous necrosis, acid fast bacilli or fungal organisims. The impression of biopsy suggested to be a case of cheilitis granulomatosa. Patient's chest x-ray and blood screening was also done. Chest xray was found normal. Other parameters of blood were normal except raised ESR. Once the case was diagnosed as Cheilitis Granulomatosa. She was given intralesional triamcinolone along with clofazimine. Swelling was reduced to large extent. But as soon as when drugs were discontinued. Swelling started to reappear. She was also given doxycline (50mg) for 3 months but not of great relief. Patient was also treated with DAPSONE, but treatment was stopped due to development of anemia. The differential diagnosis was also taken into account, that included Cheilitis Granulomatosa, Angioedema, Cheilitis glandularis, Neurofibroma, Exfoliative cheilitis, Sarcoidosis, Crohn's disease, Tuberculosis, Hemangioma, Lymphangioma, and other such conditions. The chief complaint, history, clinical examination and subsequent investigations led us in arriving at a final diagnosis of Cheilitis granulomatosa of the lower lip.

DISCUSSION

The diagnosis of cheilitis granulomatosa is based on clinical evaluation and verified by means of biopsy.Cheilitis granulomatosa is a chronic swelling of the lip due to granulomatous inflammation. It is a monosymptomatic form or an incomplete variant of MRS which is a triad of recurrent orofacial swelling, relapsing facial paralysis and fissuring of the tongue (lingua plicata). Presentation of complete MRS with all three elements of the triad in a single patient is rare, being reported in only 10-20% of cases, and bilateral facial palsy in MRS is even rarer. (Muhammed *et al*., 2004) In approximately 40% of cases, CG is the presenting sign of MRS, with subsequent development of the neurologic signs. (Muhammed *et al*., 2004) The presence of CG without lingua plicata or facial palsies also is called Miescher syndrome or Miescher's Cheilitis. (Worsaae *et al*., 1982; van der Waal *et al*., 2001) In 1985, Wiesenfield (1985) introduced the concept of Orofacial granulomatosis (OFG). Today both CG and MRS are considered subsets of OFG. The etiology of CG continues to remain a mystery. Reports suggest that some casesmay demonstrate an autosomal dominant inheritance pattern, with the responsible gene mapping to chromosome 9 p11. (Scully *et al*., 2010) Other authors have proposed a wide range of causes including allergic reactions, chronic infectious odontogenic foci, autoimmune mechanism, as an association with Crohn's disease and sarcoidosis or even as oral manifestation of systemic diseases. (Muellegger *et al*., 2000) The age of onset is usually young adulthood, second decade of life. In patients presenting with CG it is important to perform an appropriate evaluation, which includes a chest radiograph and a Mantoux test to exclude other etiologies of granulomatous disease, such as sarcoidosis or Mycobacterium infection. Gastrointestinal tract endoscopy and radiography may be used to exclude Crohn's disease. Patch tests have implicated cobalt and the food additives cinnamaldehyde and benzoates in the pathogenesis of CG. (Wiesenfeld *et al*., 1985)

Granulomatous cheilitis should be considered in children as well according to Oliver, *et al*. (2008) who reported Granulomatous Cheilitis in an eight-year-old girl, which persisted for more than one year. Zimmer, (1992) in their analyses of 42 patients and review of 220 cases they showed female predilection and a wide range of onset with a mean of 33.8 years.

Treatment and management

Management of cheilitis granulomatosa depends on accurate diagnosis of the condition and identification of any underlying factors. Patients without dental infections who present with clinical features suggestive of CG should be questioned regarding the presence of systemic signs and symptoms of crohn's disease, sarcoidosis or a history of angioedema. In the presence of positive findings, the patient should be considered for proper medical evaluation. Various treatments for cheilitis granulomatosa have been reported, including antibiotics like tetracycline and clofazimine oral and intralesional steroids, and surgical resection. Rapid improvement and/or complete resolution after dental treatment have been reported. Surgical intervention and radiation have been proposed in the management of CG in cases of severe disfigurement and post-surgical relapses are common. Some believe that intralesional triamcinolone (TAC) combined with dapsone yields the best result, whereas others report success using intralesional TAC alone; incidentally TAC has also been successfully combined with clofazimine.

Conclusion

The diagnosis of cheilitis is made by proper patients history, clinical features and histopathological examination. Intralesional triamcinolone combined with clofazimine is very much effective in handling cases of CG. Surgical option should be think of to combat cosmetic challenges.

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