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Melkersson – Rosenthal Syndrome- A Case Report of a Rare Disease with the Classic Triad

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ABSTRACT

An interrelationship between periodontal disease and systemic health has been discussed for long time. Periodontal disease can be secondary manifestations of systemic diseases. The Melkersson Rosenthal syndrome is a neuro-mucocutaneous disease. This syndrome consists of triad of intermittent facial paralysis, recurrent facial edema and lingual plicata. The cause of the syndrome is yet unknown. This paper discusses the clinical features and management of this unusual disorder.

Keywords: Melkersson Rosenthal syndrome, Cheilitis granulamatosa, lingual plicata.

INTRODUCTION

The Melkersson Rosenthal syndrome also called Miescher Rosenthal syndrome is a rare neuro-mucocutaneous disease of unknown etiology. One study estimated its incidence at 0.08% of the general population.[1] This syndrome consists of recurrent swelling of lip or face, intermittent facial paralysis and fissured tongue.[2] Melkersson in 1928 suggested a relationship between facial paralysis and swelling of the face and in 1931 he added lingual plicata to the clinical manifestations. The complete syndrome is very rare; monosymptomatic or sequential involvement is more common.

In 1945 Miescher described "Cheilitis granulamatosa'' in several of these patients. It is characterized by persistent idiopathic swelling of the lip due to granulomatous inflammation. It is thought to be a subset of orofacial granulomatosa and it is a monosymptomatic presentation of Miescher Chelitis. However, most investigators believe that cheilitis granulomatosa and Melkersson Rosenthal syndrome are same disorder (Hornstein, 1970; Gorlin et al 1976). In this case report, a patient with MRS who had applied with complaints related to recurring swelling of face is presented. This case is unique because the patient reported with all the traid including recurrent swelling of lip and face, intermittent facial paralysis due to adenoma of pituitary gland and fissured tongue. Approximately 300 cases have been reported by national organization for rare disorders, but the condition may be underdiagnosed because facial palsy is not always present.

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CASE REPORT

A 32-year-old female patient reported to the department of periodontology, Buddha Institute of Dental Sciences & Hospital, Patna complaining of swelling of face and lip for two years and bleeding gums. She also mentioned about the multiple ulcers occur every month. So, she could not brush properly while that period. The patient was apparently healthy, well oriented, was born full term & delivered normally. Her medical history revealed:

- "Cheilitis granulomatosa'' confirmed from biopsy report and Hypothyroidism.
- Facial paralysis, benign pituitary adenoma and had undergone surgery 1 year back in AIIMS, Delhi.
- Taking betamethasone injection (7mg/ml) i.v once in a month for granulomatous cheilitis and thyrox (50 mg) for hypothyroidism.

Clinical Examination

- On extra oral examination- generalized nontender, nonpitting edema of face and upper and lower lip was present. Facial nerve function was not intact on left side of face and face used to tilt to left side while talking and chewing. [Figure 1] (Swelling of face).
- On intra oral examination- generalized grade II gingival enlargement involving both facial and palatal gingiva both with gingival recession w.r.t 32 and 33 were seen. Gingiva was generalized red in color and edematous in consistency. Generalized attrition and erosion were present. Periodontal pocket depth of 6-7mm was present with placated tongue. Grade II mobility was also present w.r.t 31, 32, 41 and 42. [Figure 2a, 2b] (2a- Placated

tongue). (2b- Gingival enlargement, recession, erosion, attrition with change in color)

Investigations

Investigations that were carried out in this case included an OPG (Ortho Pantomograph). The OPG revealed severe generalized horizontal bone loss with vertical bone loss pattern wrt 15-17, 26-27 and 27-28. [Figure 3] (OPG done)

Laboratory values, including complete blood cell count, blood sugar

(Fasting & PP), bleeding time, clotting time, prothrombin time, hemoglobin, T3, T4 and TSH and biopsy was done for histopathologic findings. Histopathologic evidence is not necessary for the diagnosis of Melkersson-Rosenthal syndrome8. The results of basic laboratory tests were normal and histopathology report showed chronic inflammatory cells and mild granuloma in lamina propria with dilated & engorged blood vessels. [Figure 4] (Blood investigation and histopathology done with H& E stain)

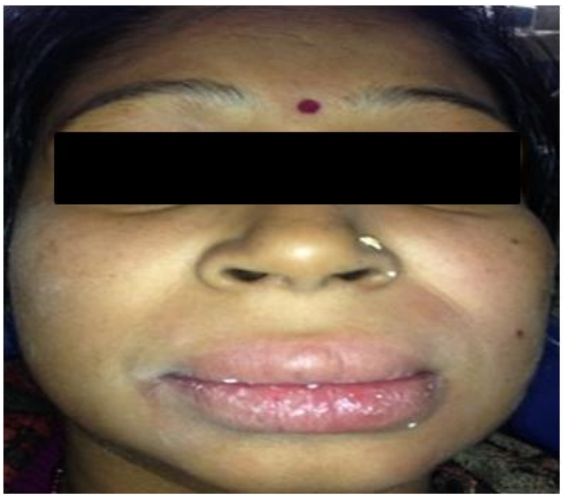

Figure 1: Swelling of face

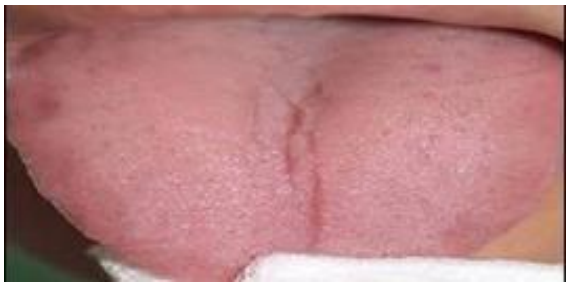

Figure 2a: Placated tongue

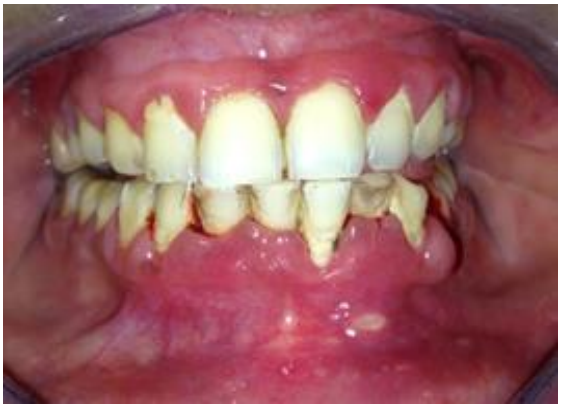

Figure 2b: Gingival enlargement, recession, erosion, attrition with change in color

Figure 3: OPG done

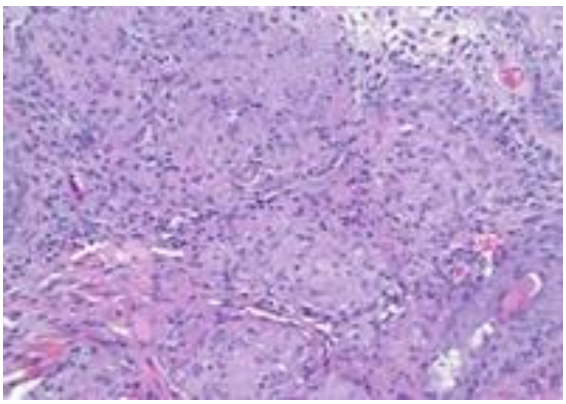

Figure 4: Histopathology done with H&E stain

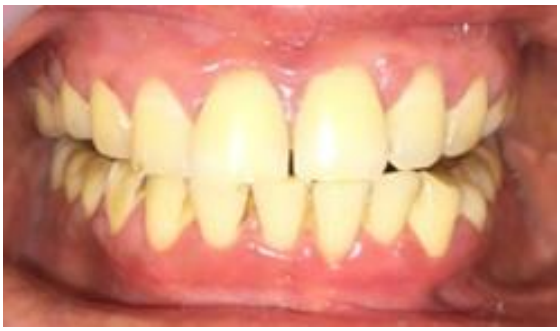

Figure 5: Post– operative picture

Periodontal Management

The complete treatment procedure was explained to the patient, a medical consent was obtained from concerned physician and a written informed consent was obtained from the patient.

- A thorough scaling & root planning were done carefully to remove any local irritating factors that may have been responsible for the superimposed gingival inflammation.
- The patient was given oral hygiene instructions and triamcinolone gel (oralone) twice daily for oral ulcers.
- 21 days later patient came with reduced plaque levels, enough to advocate phase II therapy.
- As there was systemic disease associated, a quadrant wise surgical procedure was performed.
- Antibiotic coverage (Amoxicillin 1000 mg/stat, 1 hr prior to surgery) was given. Firstly, external bevel gingivectomy was done w.r.t lower right and left anterior sextants. This was done to obtain proper physiological contour.
- Conventional flap therapy was performed for all the other arches.
- There was normal level of intraoperative bleeding associated with affected site.
- Immediately after surgical procedure, patient was advised to continue antibiotic-analgesic regimen (Tab cifran 500mg, twice daily + Tab megaflam AC (425mg), twice daily for next 5 days). 0.2% chlorhexidine mouthwash, 10ml twice daily for 1-minute, half an hour after brushing.
- Post-operative instructions were given to the patients.

  6 months post-operative visit of the patient revealed no signs of recurrent enlargement and pocket depth was also reduced. [Figure 5] (Post–operative picture).

DISCUSSION

The Melkersson Rosenthal syndrome also called Miescher Rosenthal syndrome. Melkersson in 1928 suggested a relationship between facial paralysis and swelling of the face and in 1931 he added lingual plicata to the clinical manifestations.

Cheilitis granulomatosa is the most frequent clinical manifestation in patients with MRS. Histopathology may help to exclude any other conditions but it has no need for the diagnosis.

In the present case, a careful history was taken, all the parameters were recorded, and all the investigations were done carefully for proper diagnosis and management of disease. The treatment of Melkersson Rosenthal syndrome is variable & depends on the nature and clinical severity of the disease. Oral or intralesional steroids remain a mainstay of treatment regimens and its potent anti-inflammatory activity is associated with clinical and histological improvement.[9] Anti-inflammatory and immunomodulatory activities of some antibiotic agents may explain why they have been successfully used for treating cheilitis granulomatosa. The antibiotics that have gained more recent prominence include minocycline (100 mg daily), roxithromycin (150-300 mg daily), and metronidazole (750-1000 mg daily). Infliximab (3-5 mg/kg) also has same role as antibiotics.[10,11]

Methotrexate reduces the proliferation of immune cell lines and was used effectively in doses of 5-10 mg given orally at weekly intervals.[10-12]

A good oral hygiene, intermittent use of an anti-inflammatory and alkali solutions may be of benefit in these cases.[13] This study also concluded that good oral hygiene of the patient showed no further signs of periodontal disease.

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CONCLUSION

Melkersson-Rosenthal syndrome is a rare disorder with the classic triad. The patient displayed the classic triad like orofacial swelling, plicated tongue and facial palsy. The patient was afflicted because she couldn't maintain oral hygiene properly. After periodontal treatment and proper oral hygiene instructions, she had no sign of periodontal disease after 6 months.

REFERENCES

1. Hakim M E L. J Oral Maxillofac Surg 2004.
2. Hathiram B T, Grewal D S, Walvekar R, Dwivedi A, Kumar L & Mohorikar A. Melkersson Rosenthal syndrome As A Rare Cause OF Facial Palsy-A Case Report. Indian J of Otolaryngology and Head and Neck Surgery. 2000.
3. James W D. Cheilitis Granulomatosa. American Academy of Dermatology, Society for Investigative Dermatology. 2017.
4. Hornstein OP. Melkersson Rosenthal syndrome. A neuro-muco-cutaneous disease of complex origin. Curr Probl Dermatol. 1973.
5. White A, Nunes C & Escudier M. Improvement in orofacial..
6. Ketabchi S, Massi D & Ficarra G. Expression of protease-activated receptor granulomatosis on a cinnamon and benzoate-free diet. Inflamm Bowel Dis. 2006.
7. 1 and 2 in orofacial granulomatosis. Oral Dis. 2007.
8. Smeets E, Fryns JP & Berghe V D H. Melkersson Rosenthal syndrome and de novo autosomal t (9; 21) (p11; p11) translocation. Clin Genet. 1994.
9. Feng S, Yin J, Li J, Song Z & Zhao G. Melkersson Rosenthal syndrome: a retrospective study of 44 patients. Acta Otolaryngol. 2014.
10. Banks T & Gada S. A comprehensive review of current treatments for granulomatous cheilitis. Br J Dermatol. 2012.
11. Tonkovic Capin V, Galbraith SS, Rogers RS 3rd, Binion DG & Yancey KB. Cutaneous Crohn's disease mimicking Melkersson Rosenthal syndrome: treatment with methotrexate. J Eur Acad Dermatol Venereol. 2006.
12. Ratzinger G, Sepp N, Vogetseder W & Tilg H. Cheilitis granulomatosa and Melkersson Rosenthal syndrome: evaluation of gastrointestinal involvement and therapeutic

regimens in a series of 14 patients. J Eur Acad Dermatol Venereol. 2014.
13. Berry O, Barry J, Langan S, Murphy M, Fitzgibbon J & Lyons JF. Treatment of granulomatous cheilitis with infliximab. Arch Dermatol. 2005.
14. Nisa L & Giger R. Lingua plicata. CMAJ. 2012.

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