ORIGINAL RESEARCH PAPER

Volume-8 | Issue-3 | March-2019 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

MELKERSSON ROSENTHAL SYNDROME- A CLINICAL REVIEW

Neurology

Dr Abhas Kumar DNB Neurology, Institute of Neurosciences, Indraprastha Apollo Hospitals – 110076

Pushpendra N Renjen* Sr. Consultant Neurologist & Academic Coordinator, Institute of Neurosciences, Indraprastha Apollo Hospitals *Corresponding Author

Dr. Dinesh Chaudhari Clinical Associate, Institute of Neurosciences, Indraprastha Apollo Hospitals – 110076

ABSTRACT

Melkersson-Rosenthal Syndrome (MRS) is a rare disorder and consists of a triad of recurrent orofacial edema, recurrent facial palsy and fissured tongue. All aspects of the classical triad may not be observed at the same time, as they may appear at different times. The most common symptom is orofacial edema. The etiology and mechanisms of the syndrome have not been fully elucidated. We report a case of 25-year-old male, with history of recurrent left facial palsy. Clinical examinations at presentation revealed classical triad of MRS.

KEYWORDS

Melkersson-Rosenthal Syndrome; Orofacial edema; Recurrent facial palsy; Fissured tongue.

INTRODUCTION

Melkersson Rosenthal Syndrome (MRS) consists of the clinical triad of recurring facial nerve palsy, swelling of lips and fissured tongue. (1) Usually all the components of the triad are not seen together. Oligo-symptomatic or mono-symptomatic form is the most common forms seen in day to day practice. Orofacial edema, involving lower lip or both lips, is the most common finding. (2) A fissured tongue is seen in 20-40% of those affected and may be present since birth. Facial palsy occurs in about 30- 35% of those affected.(2,3) Females are affected more than males.(2) Recurrent episodes may lead to worsening and persistent swelling, which may become permanent.(3) Melkersson Rosenthal Syndrome is inherited in autosomal dominant fashion, but a consistent genetic cause has not been found. It is possible that more than one gene is responsible for MRS, and/or that environmental "triggers" may contribute to causing the syndrome in some genetically predisposed individuals.(2) Etiolopathogenesis of Melkersson Rosenthal Syndrome is unclear, various factors act as triggers such as infections, genetic predisposition, immune deficiency, food intolerance and stress.(4) In this case report we report a patient who presented with classical triad of Melkersson Rosenthal Syndrome, which itself is a very rare finding in clinical neurology.

CASE REPORT

25 years-old male patient, with no systemic diseases, presented fourth episode of left facial palsy. Patient's history revealed that he had suffered three episodes facial paralysis on the left side of his face over the past 2years which recovered completely. During examination, left LMN facial palsy (Figure 1a and 1b) was observed along with swelling of the lower lip was also observed. The patient reported that the swelling in the upper lip had been observed in the past (Figure 2). Oral examination revealed that the patient had fissured tongue (Figure 3a and 3b). Patient was evaluated with CT temporal bone and MRI brain with MRI of salivary gland, both of which had revealed no evidence of temporal bone or mastoid destruction or salivary gland tumour. Serum ACE levels were normal (38 nmol/ml/min). The patient was managed with steroids, antihistaminics and other supportive care.

Figure 1 a

Figure 1b

Figure 2

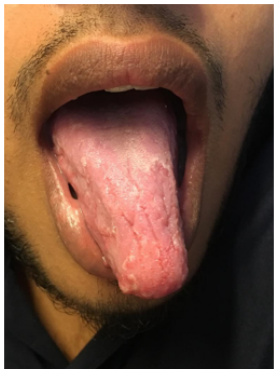

Figure 3a

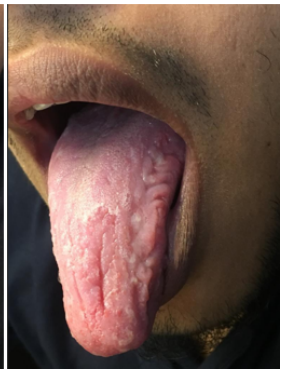

Figure 3b

DISCUSSION

Melkersson Rosenthal syndrome is a rare neurological disorder characterized by recurrent, long lasting swelling of the face, particularly one or both lips (granulomatous cheilitis), facial muscle weakness (palsy) and a fissured tongue. Classical triad of MRS can only be observed in 8% to 18% of the patients, which makes it difficult to diagnose this condition. (5,6) Melkersson Rosenthal syndrome is thought to be caused by genetic factors in some cases because families have been described in which multiple members are affected. (7) Melkersson Rosenthal syndrome is sometimes a symptom of another condition such as Crohn's disease or sarcoidosis.

The most common finding of Melkersson Rosenthal syndrome is painless and non-pitting orofacial edema which is most frequently seen on lips.(5) Melkersson Rosenthal syndrome resembles angioedema at times, but it can be differentiated with the lack of response to antihistaminic drugs, tendency to last longer and by causing fibrosis in the involved tissues.(8) The second characteristic finding of Melkersson Rosenthal syndrome is facial paralysis which can be observed in 30% to 35% of the patients (9). Paralysis can be

transient, or can become permanent in time. It can be unilateral as well as bilateral. The paralysis has been attributed to either to the edema on the nerve passing through the facial canal or the infiltration of the nerve (10, 11). The third and least specific finding of Melkersson Rosenthal syndrome is the fissured tongue. Since fissured tongue is a frequent finding among masses, it is of lesser importance in the diagnosis (12). Melkersson Rosenthal syndrome is diagnosed by physical findings and history, hence is a clinical diagnosis. A biopsy of the lips may be necessary to confirm the diagnosis in some cases to rule out granulomatous chelitis. Specialized laboratory studies and procedures may be recommended to rule out Crohn's disease and sarcoidosis.

Most of the manifestations resolve spontaneously at times. Despite controlling the manifestations with medications, it can recur in the future. (13) Edema in the orofacial area can be cured with the systemic use of steroids or intra-lesional corticosteroid administration. (14,15) Sometimes surgical intervention might be required for decompression of facial nerve. Role of immunosuppressive agents and adalimumab needs to be evaluated further in the management of this disease. (16)

CONCLUSION

Melkersson Rosenthal syndrome is a very rare disease and is diagnosed clinically based on its triad of orofacial edema, recurrent facial palsy and fissured tongue. Although all the three components might not be evident at the same time and hence can be misdiagnosed. Melkersson Rosenthal syndrome is an important cause of recurrent facial palsy and must be considered while evaluating patients with recurrent facial palsy with negative evaluation of local anatomical and central causes.

REFERENCES

1. Ang KL, Jones NS. Melkersson-rosenthal syndrome. J Laryngol Otol. 2002;116(5):386–388.
2. Pei Y, Beaman GM, Mansfield D, Clayton-Smith J, Stewart M, Newman WG. Clinical and genetic heterogeneity in Melkersson-Rosenthal Syndrome. Eur J Med Genet. September 11, 2018; [Epub ahead of print]:https://www.sciencedirect.com/ science/ article/ pii/S176972121830394X.
3. Melkersson-Rosenthal Syndrome Information Page. National Institute of Neurological Disorders and Stroke (NINDS). August 9, 2018; https://www.ninds.nih.gov/Disorders/ All- Disorders/ Melkersson-Rosenthal-Syndrome-Information-Page.
4. Basman A, Gumusok M, Degerli S, Kaya M, Toraman Alkurt M. Melkersson-rosenthal syndrome: a case report. J Istanb Univ Fac Dent. 2017;51(1):42-45. Published 2017 Jan 2. doi:10.17096/jiufd.96279.
5. Balevi B. Melkersson-rosenthal syndrome: Review of the literature and case report of a 10-year misdiagnosis. Quintessence Int. 1997;28(4):265–269.
6. Zimmer WM, Rogers RS, Reeve CM, Sheridan PJ. Orofacial manifestations of melkersson-rosenthal syndrome. A study of 42 patients and review of 220 cases from the literature. Oral Surg Oral Med Oral Pathol. 1992;74(5):610–619. doi: 10.1016/0030-4220(92)90354-S.
7. Ziem PE, Pfrommer C, Goerdt S, Orfanos CE, Blume-Peytavi U. Melkersson-rosenthal syndrome in childhood: A challenge in differential diagnosis and treatment. Br J Dermatol. 2000;143(4):860–863. doi: 10.1046/j.1365-2133.2000.03791.
8. Alexander RW, James RB. Melkersson-rosenthal syndrome: Review of literature and report of case. J Oral Surg. 1972;30(8):599–604.
9. Gerressen M, Ghassemi A, Stockbrink G, Riediger D, Zadeh MD. Melkersson-rosenthal syndrome: Case report of a 30-year misdiagnosis. J Oral Maxillofac Surg. 2005;63(7):1035–1039. doi: 10.1016/j.joms.2005.03.021.
10. Sciubba JJ, Said-Al-Naief N. Orofacial granulomatosis: Presentation, pathology and management of 13 cases. J Oral Pathol Med. 2003;32(10):576–585. doi: 10.1034/j.1600-0714. 2003.t01-1-00056.x.
11. Leao JC, Hodgson T, Scully C, Porter S. Review article: Orofacial granulomatosis. Aliment Pharmacol Ther. 2004;20(10):1019–1027. doi: 10.1111/j.1365-2036.2004. 02205.
12. van der Waal RI, Schulten EA, van de Scheur MR, Wauters IM, Starink TM, van der Waal I. Cheilitis granulomatosa. J Eur Acad Dermatol Venereol. 2001;15(6):519–523. doi: 10.1046/j.1468-3083.2001.00353.
13. Kemal O, Özgürsoy OB, Dursun G, Tulunay O. Melkersson-rosenthal sendromu klinik ve patolojik bulgular ve tedavi yaklaşımları Turkiye Klinikleri J Med Sci. 2007;27(1):128–131.
14. Cerimele D, Serri F. Intralesional injection of triamcinolone in the treatment of cheilitis granulomatosa. Arch Dermatol. 1965;92(6):695–696. doi: 10.1001/archderm.1965. 01600180087017.
15. Krutchkoff D, James R. Cheilitis granulomatosa. Successful treatment with combined local triamcinolone injections and surgery. Arch Dermatol. 1978;114(8):1203–1206. doi: 10.1001/archderm.1978.01640200057015.
16. de Moll EH, Lebwohl MG. Melkersson-Rosenthal syndrome successfully treated with adalimumab. Cutis. 2018 Feb;101(2):122-124. PubMed PMID: 29554155.