

Melkerssons-Rosenthal syndrome: A case report and review of the literature

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Abstract

Melkerssons-Rosenthal syndrome is a clinical entity identified by the presence of the triad of recurrent facial paralysis, recurrent often permanent (labial) oedema, and to a lesser extent the placation of the tongue. The striking event is that of recurrent lower motor facial paresis. This should arouse the search for the other components of the triad which is not always complete. The major worry of the patients is however the facial paresis/oedema and the attendant inability to close the eyes which might sometimes get infected. Hallmark of management remains physical therapy and corticosteroid and/or antiviral agents. A high index of suspicion is required to make the diagnosis of Melkerssons-Rosenthal syndrome.

Key words: Melkerssons-Rosenthal, Syndrome, Triad, Index of Suspicion

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Introduction

Melkerssons-Rosenthal syndrome is an uncommon neurocutaneous disorder which often present with recurrent episodes of orofacial swelling, lower motor facial palsy, and fissured tongue. It has also been regarded as a non-caseating granulomatous disease of unclear etiology.^[1-3,5]

Presentations may be partial or complete. Many at times the main feature at presentation will be just facial edema involving the eyelids.^[4,5] Etiology and pathogenesis of MRS have not been fully determined. Several factors, such as infection, autoimmunity, neurotropic factors, atopy, and hypersensitivity to food additives have been implicated in the pathogenesis, but none of them are clearly proven.^[6-9]

Case Report

Melkerssons-Rosenthal syndrome is defined by the triad of recurrent facial paralysis, recurrent often permanent (labial) oedema, and rarely placation of the tongue.

This 35-year-old woman whose consent was sorted, presented with four episodes of recurrent right lower motor facioparesis, right faciolabial oedema, with placation noticed over her tongue [Figures 1 and 2].

On the first two occasions she was treated with oral prednisolone and physical therapy with significant clinical improvement with each treatment only to recur.

On presentation, at the last and forth occasion she was treated with acyclovir 400 mg five times daily for 10 days in addition to prednisolone 60mg daily for 6 days which was subsequently tapered off over the next 6 days.

She has since remained asymptomatic with no recurrence in the last 24 months. We still keep a monthly contact with her.

Ethical approval for this case report was obtained from the ethical committee of the State Hospital, Abeokuta, Nigeria. Patients' informed consent of the use of her picture for this article was also obtained.

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Figure 1: Melkerssons-Rosenthal syndrome

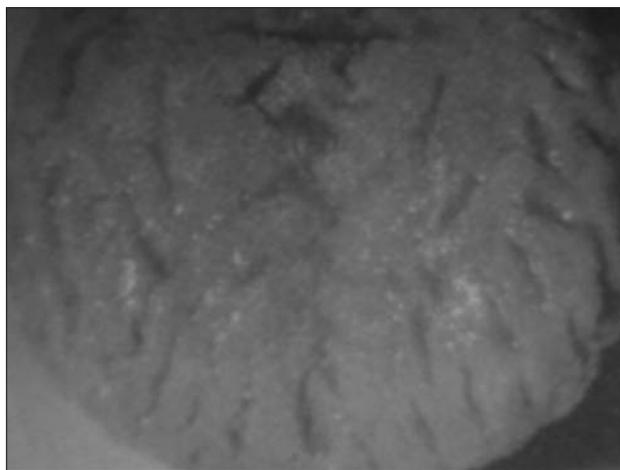


Figure 2: Placation of tongue

Another possible mechanism is the vasomotor disturbances of both the vasa nervorum and the small arterioles of the subcutaneous tissues in response to unspecified stimuli in predisposed persons. An autosomal dominant inheritance with variable expression has been proposed in some cases of MRS

If the etiology is unclear, the diagnosis and course of treatment are not well defined.^[9]

Various modalities of treatment have been tried, among which include corticosteroid, doxycycline, clofazimine, and thalidomide among others.^[10-11] A familial tendency has been reported.^[11]

The presented case, after three clear episodes of about 2-3 months intervals has remained quiet after the forth for over 2 years now.

References

1. Balevi B. Melkersson-Rosenthal Syndrome: Review of the literature and case report of a 10 year misdiagnosis. *Quintessence Int* 1997;28:265-9.
2. Green RM, Rogers RS. Melkersson-Rosenthal Syndrome: A review of

- 36 patients'. *J Am Acad Dermatol* 1989;21:1263-70.
3. Glickman LT, Gruss JS, Birt BD, Kohli-Dang N. The surgical management of Melkersson-Rosenthal Syndrome. *Plast Reconstr Surg* 1992;89:815-21.
4. Ghorbel IB, SioudDhrifA, Lamoum, Trabelsi S, Habib Houman M. Melkersson-Rosenthal syndrome. Report of five cases and review of the Literature. *Tunis Med* 2006;84:816-20.
5. Marques C, Machado A, Baptista AP. Marcrochelitis and Melkersson-Rosenthal Syndrome. Report of five cases and review of the literature *Tunis Med*, 2006 Dec; 84:816-20.
6. Marques C, Machado A, Baptista AP. Marcrochelitis and Melkers son-Rosenthal syndrome. Review of 19 cases. *Acta Med Port* 1994;7:533-40.
7. Hintjens J, Daems L, Boss uyt M. Melkersson Rosenthal syndrome. Review of the literature. *Acta Stomatol Belg* 1994;91:143 -54.
8. Cousin F, Grezard P, Berard F, Perrot H. Melkersson-Rosenthal syndrome. *Ann Med interne (Paris)* 1998;149:495-501.
9. Medeiros M Jr, Araujo MI, Guimarães NS, Freitas LA, Silva TM, Carvalho EM. Therapeutic response to thalidomide in Melkersson-Rosenthal syndrome: A case report. *Ann Allergy Asthma Immunol* 2002;88:421-4.
10. Melkersson-Rosenthal syndrome: A case report. *Ann Allergy Asthma Immunol* 2002;88:421-4.
11. Safa G, Joly P, Boullie MC, Thomine E, Lauret P. Melkers son-Ros enthal syndrome: Treated by Thalidomide: 2 Cases. *Am Dermatol Venereol* 1995;122:609-11.

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