

Outcomes of Recurrent Facial Palsy in Melkersson Rosenthal Syndrome

Annals of Otolaryngology & Laryngology
2015, Vol. 124(3) 232–234
© The Author(s) 2014
Reprints and permissions:
sagepub.com/journalsPermissions.nav
DOI: 10.1177/0003489414551981
aor.sagepub.com



Junying Wang, MM¹, Peizhong Li, MD¹, Xin Jin, BM¹, Yu Xu, BM¹,
and Xin Zhang, BM¹

Abstract

Objective: The objective of the study was to investigate outcomes of recurrent facial palsy in Melkersson Rosenthal syndrome (MRS).

Methods: We carried out a prospective investigation on patients who had recurrent facial palsy in MRS and visited an outpatient department of otolaryngology from January 2002 to January 2012, focusing on their facial nerve outcomes. They were followed up for 12 months. The patients who visited our department due to recurrent Bell's palsy during the same period were selected as controls. Oral prednisolone alone was given to each patient.

Results: A total of 22 patients with recurrent facial palsy in MRS and 73 patients with recurrent Bell's palsy were included in the study. Twelve months later, only 3 of 22 patients (13.6%) who had recurrent facial palsy in MRS recovered to House-Brackmann grade I, whereas 52 of 73 patients (71.2%) who had recurrent Bell's palsy recovered completely. The difference was statistically significant ($P < .01$).

Conclusion: Compared to recurrent Bell's palsy, recurrent facial palsy in MRS had much worse prognosis.

Keywords

Bell's palsy, Melkersson Rosenthal syndrome, recurrent facial palsy

Introduction

Melkersson Rosenthal syndrome (MRS) is featured by recurrent orofacial edema, recurrent peripheral facial palsy, and lingua plicata.¹ It is a rare disease, and the etiology is still unclear. Viral infection, allergic factors, and hereditary origin have been proposed, but there is no persuasive evidence.^{2–4} Facial palsy in MRS is quite rare compared to Bell's palsy. According to a 25-year prospective study of peripheral facial palsy etiology,⁵ the incidence of MRS with facial palsy is 0.36 in 100 000 patients per year, whereas that of Bell's palsy is 20 to 30 in 100 000 patients per year.

Patients with MRS usually visit a department of otolaryngology due to facial palsy. It is reported that 75% of MRS patients with facial palsy have at least 2 episodes of facial palsy,⁶ indicating that recurrent facial palsy is quite common in MRS. There have been several reports discussing outcomes of recurrent Bell's palsy,^{7–10} but the study focusing on outcomes of recurrent facial palsy in MRS is defective, to our knowledge, possibly due to its rarity. The clinical practice was impressive in that recurrent facial palsy in MRS had worse prognosis than recurrent Bell's palsy. To ascertain whether recurrent facial palsy in MRS really had worse recovery, we performed the prospective study in which 22 patients with recurrent facial palsy in MRS and 73 patients with recurrent Bell's palsy were involved.

Materials and Methods

The study covered the patients with recurrent facial palsy in MRS and those with recurrent Bell's palsy who visited the outpatient department of otolaryngology between January 2002 and January 2012. Facial palsy recurrence may be ipsilateral or alternating. The patients who had at least 2 symptoms of recurrent orofacial edema, recurrent peripheral facial palsy, and lingua plicata were clinically diagnosed as having MRS, and recurrent Bell's palsy was differentiated from recurrent facial palsy in MRS mainly according to the absence of recurrent orofacial edema and lingua plicata. Patients who met any of the following were excluded from the study: facial nerve tumors, otitis media and acoustic neuromas, malignant hypertension, and Ramsay Hunt syndrome. Thorough neurological examination, high-resolution computed tomography of temporal

¹Department of ENT, Huai'an First People's Hospital, Nanjing Medical University, Huai'an, China

Corresponding Author:

Junying Wang, MM, Department of ENT, Huai'an First People's Hospital, Nanjing Medical University, No. 6 Beijing Road West, Huai'an 223300, Jiangsu Province, P.R.C.
Email: wangjy4203@163.com

Table 1. Characteristics of Patients With Recurrent Facial Palsy in Melkersson Rosenthal Syndrome (MRS) and Recurrent Bell's Palsy.

Group	Recurrent Facial Palsy in MRS	Recurrent Bell's Palsy	P Value
Age, y, mean \pm SD	21.3 \pm 12.4	46.2 \pm 18.2	< .05
Male/female	10/12	38/35	> .05
Ipsilateral/contralateral	14/8	40/33	> .05
Site, left/right	13/9	39/34	> .05
First/second or more recurrence	12/10	38/35	> .05
Grade, mean \pm SD	4.7 \pm 1.1	4.1 \pm 0.7	> .05

bone, and magnetic resonance imaging were used for differential diagnosis.

Concerning treatment, oral prednisolone alone (1 mg/kg/d for 1 week, and then reduced by 10 mg every 2 days) was offered to each patient with facial palsy in MRS or Bell's palsy within 1 week after facial palsy. They were followed up for 12 months, and the House-Brackmann (HB) grading scale was introduced to assess facial nerve function.¹¹ The clinical characteristics of the patients were recorded and analyzed. Chi-square test and *t* test were used in statistical analysis, and SPSS 19.0 software was involved.

Informed consent was obtained from each patient, and the study passed approval of the ethics committee in our hospital.

Results

A total of 22 patients with recurrent facial palsy in MRS and 73 patients with recurrent Bell's palsy among 1146 patients with peripheral facial palsy who conformed to the inclusion criteria were included in the study. Among 22 patients with MRS, there were 12 in triad and 10 in oligosymptomatic form. The diagnosis was clinically made in 14 patients and histologically confirmed in 8 patients.

Their clinical characteristics are listed in Table 1. The patients had a mean (SD) age of 21.3 (12.4) years, and 10 patients (45.5%) were male. Fourteen patients (63.6%) had ipsilateral recurrence of facial palsy, and 13 (59.1%) had left-side involvement. It was the first recurrence in 12 patients (54.5%). Mean (SD) HB grade of facial nerve function before treatment was 4.7 (1.1), ranging from grade II to grade VI, with grade VI in 77.3% of patients.

In contrast, the mean (SD) age of the patients with recurrent Bell's palsy was 46.2 (18.2) years, and 38 patients (52.1%) were male. Forty of the patients (54.8%) had ipsilateral recurrence, and 39 patients (53.4%) had left-sided facial palsy. Thirty-eight patients (52.1%) had the first recurrence. Initial mean (SD) HB grade of facial nerve was 4.1 (0.7), ranging from grade II to grade VI, with grade VI in 71.2% of patients. The 2 groups were similar in sex distribution, ipsilateral recurrence, involved site, first recurrence ratio, and initial grade ($P > .05$) (Table 1). However, the patients who had recurrent facial palsy in MRS were younger than those who had recurrent Bell's palsy ($P < .05$).

Table 2. Outcomes of Recurrent Facial Palsy in Melkersson Rosenthal Syndrome (MRS) and Recurrent Bell's Palsy.^a

Group	Complete Recovery	Incomplete Recovery	Total (Cases)
Recurrent facial palsy in MRS	3	19	22
Recurrent Bell's palsy	52	21	73
Total	55	40	95

^a $\chi^2 = 23.006, P = .000$.

Twelve months after facial palsy, only 3 of 22 patients (13.6%) who had recurrent facial palsy in MRS recovered to grade I, and 19 patients (86.4%) had sequelae of facial nerve. In contrast, 52 of 73 patients (71.2%) who complained of recurrent Bell's palsy recovered to grade I, and the remaining 21 patients (28.8%) had incomplete recovery. The difference was dramatically significant ($P < .01$).

Discussion

Facial palsy is common in MRS, but the pathogenic mechanism is still unknown. Some authors^{5,12} think that facial palsy in MRS mimics Bell's palsy, in which viral infection may be the etiological factor, but virus is not even found in Bell's palsy.¹³ Unlike Bell's palsy, in which recurrent facial palsy is present in only 8.2% of the population,⁷ the majority of patients who have facial palsy in MRS complain of facial palsy recurrence.

The prognosis of recurrent Bell's palsy is controversial. van Amstel and Devriese⁷ retrospectively analyzed 106 patients with recurrent Bell's palsy out of 1296 Bell's palsy patients during a 10-year period and found that facial nerve recovery of those patients who had ipsilateral recurrence was worse in 39.6% of patients. Similar results were reported by several other authors.^{8,14,15} However, Pitts et al¹⁶ also carried out a retrospective study on a larger series of recurrent Bell's palsy within a 9-year period and concluded that recurrent facial paralysis did not indicate a worse prognosis for recovery regardless of which side was affected, as was also verified by another prospective study that involved 280 patients.

The information about recurrent facial palsy in MRS is very limited, possibly due to its rarity. Kanerva et al⁶ mentioned outcomes of 9 patients with recurrent facial palsy in MRS, but the treatment details were not specified and, in addition, there was no control group. To find outcomes of recurrent facial palsy in MRS, we carried out a prospective study on a larger case series and set recurrent Bell's palsy as the control. In the study, 13.6% of the patients with recurrent facial palsy in MRS recovered to grade I during the 12-month follow-up, in contrast to 71.2% of the patients with recurrent Bell's palsy ($P < .01$), and 86.4% of the patients with recurrent facial palsy in MRS had sequelae of an alternate degree. The results demonstrated that recurrent facial palsy in MRS had much worse prognosis than recurrent Bell's palsy. Moreover, the study showed that recurrent facial palsy in MRS and recurrent Bell's palsy had similar sex distribution, ipsilateral recurrence, involved site, first recurrence ratio, and initial grade, although the former tended to affect younger patients.

Currently, prednisolone is the leading treatment for facial palsy in MRS in the previous literature, but whether prednisolone is really beneficial is still not determined despite its administration in our patients. In the current study, it was found that outcomes of recurrent facial palsy in MRS were still poor, although prednisolone was prescribed. Of course, we could not declare that prednisolone is not beneficial for recurrent facial palsy in MRS, since we are unclear about the natural prognosis without any treatment. It is interesting that Dai et al¹⁷ performed subtotal facial nerve decompression on 8 patients with recurrent facial palsy in MRS who showed more than 95% degeneration of facial nerve, and 3 cases (37.5%) recovered to grade I, but their study could not definitively support the benefits of surgical decompression for recurrent facial palsy due to absence of a control group.

According to our study, prognosis of recurrent facial palsy in MRS is much worse than that of recurrent Bell's palsy. Although the study is a prospective one and the case series are relatively larger, there are still some limitations. First, facial nerve function was not assessed blindly due to clinical practice. Second, electroneurography or electromyography was not performed.

Authors' Note

The authors alone are responsible for the content and writing of this article.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

References

- Greene RM, Rogers RS III. Melkersson-Rosenthal syndrome: a review of 36 patients. *J Am Acad Dermatol*. 1989;21(6):1263-1270.
- Cabrera-Gómez JA, Echazabal-Santana N, Real-González Y, et al. Hereditary Melkersson-Rosenthal syndrome and multiple sclerosis. *Mult Scler*. 2005;11:364-366.
- De Maria A, Zolezzi A, Passalacqua G, Leva M, Tarchino F, Spaggiari P. Melkersson-Rosenthal syndrome associated with parvovirus B19 viraemia and haemophagocytic lymphohistiocytosis. *Clin Exp Dermatol*. 2009;34:e623-e625.
- Herbert AA, Berg JH. Mucous membrane disorders. In: Schachner LA, Hansen RC, eds. *Pediatric Dermatology*. New York, NY: Churchill Livingstone; 1995:481-482.
- Peitersen E. Bell's palsy: the spontaneous course of 2,500 peripheral facial nerve palsies of different etiologies. *Acta Otolaryngol Suppl*. 2002;(549):4-30.
- Kanerva M, Moilanen K, Virolainen S, Vaheri A, Pitkäranta A. Melkersson-Rosenthal syndrome. *Otolaryngol Head Neck Surg*. 2008;138(2):246-251.
- van Amstel AD, Devriese PP. Clinical experiences with recurrences of Bell's palsy. *Arch Otorhinolaryngol*. 1988;245(5):302-306.
- Hallmo P, Elverland HH, Mair IW. Recurrent facial palsy. *Arch Otorhinolaryngol*. 1983;237(2):97-102.
- Ralli G, Magliulo G. Bell's palsy and its recurrences. *Arch Otorhinolaryngol*. 1988;244(6):387-390.
- Mamoli B, Neumann H, Ehrmann L. Recurrent Bell's palsy. Etiology, frequency, prognosis. *J Neurol*. 1977;216(2):119-125.
- House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg*. 1985;93(2):146-147.
- Ziem PE, Pfrommer C, Goerd 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.
- Florez RA, Lang R, Veridiano AM, et al. Intratemporal facial nerve ultrastructure in patients with idiopathic facial paralysis: viral infection evidence study. *Braz J Otorhinolaryngol*. 2010;76(5):639-642.
- Boddie HG. Recurrent Bell's palsy. *J Laryngol Otol*. 1972;86:117-120.
- Doshi J, Irving R. Recurrent facial nerve palsy: the role of surgery. *J Laryngol Otol*. 2010;124(11):1202-1204.
- Pitts DB, Adour KK, Hilsinger RL Jr. Recurrent Bell's palsy: analysis of 140 patients. *Laryngoscope*. 1988;98(5):535-540.
- Dai C, Li J, Yang S, et al. Subtotal facial nerve decompression for recurrent facial palsy in Melkersson Rosenthal syndrome. *Acta Otolaryngol*. 2014;134(4):425-428.