INTRODUCTION

Melkersson-Rosenthal syndrome (MRS) is a rare neuro-muco-cutaneous [5] granulomatous disorder of unknown etiology, characterized by the triad of facial palsy, lingua plicata (fissured tongue) and orofacial edema. [1-4] The association of edema and facial palsy was first described by Melkersson in 1928. [1, 5-8] Rosenthal added lingua plicata as an additional feature in 1931. [1, 5-8] This syndrome usually has a recurrent and progressive course [7] and unclear pathophysiology. [3, 6] The complete triad is present in a minority of the patients; [2, 9] The triad symptoms/findings can be simultaneous or present at different times. [2, 8] Although MRS is a clinical syndrome, with no

RESULTS

Twenty-one patients were identified from 1971 to 2010. The age of presentation ranged from 22 to 67 years (mean 44.1). Seven (33.3%) were male and 14 (66.7%) were female. All (100%) patients had facial paralysis. Fourteen (66.7%) patients who initially presented with unilateral paralysis subsequently developed metachronous contralateral paralysis (alternating unilateral facial paralysis). One (4.7%) patient had simultaneous bilateral facial paralysis. The number of episodes per patient ranged from 1 to 8 (mean 3.1). The patient with most episodes of facial paralysis had 4 on the left and 4 on the right (metachronous). The age at first incidence of facial paralysis ranged from 2 to 60 years (mean 34.4, median of 39). The mean interval between episodes was 4.7 years (range 0-30, median 3). Six (28.5%) of the patients reported a family history of MRS.

REFERENCES

1. Visinescu, L.M. and D.A. Karmann. The Melkersson-Rosenthal syndrome. Facial Nerve Centre, University of Pittsburgh. Pittsburgh. Each patient’s data was reviewed, extracted, and de-identified in accordance with institutional protocols. The following variables were recorded and analyzed: age, sex, number of episodes of facial paralysis, laterality, House-Brackmann score, lingua plicata, facial edema, facial paresthesia, history of diabetes, migraine/headache, tinnitus, dizziness, dysgeusia, surgical treatment and family history.

CONCLUSIONS

Melkersson-Rosenthal Syndrome is a rare disease of unknown pathogenesis in which oligosymptomatic forms predominate. Patients with this disease may present to different specialties complaining of different symptoms, and frequently, not all the classic features of the triad will be present. In our series of facial paralysis patients diagnosed with Melkersson-Rosenthal Syndrome, a higher proportion had the full triad of symptoms than has been previously reported in the literature.

Most (76.2%) of the patients in our group had lingua plicata, which is higher than previously reported, [5, 6, 8, 9] including the series by Greene et al. [50% to 60%]. [4]

It is important to note that most of our patients' (90.5%) complained of facial pain, and approximately one-third (38.1%) of facial paresthesia, which may be related to fifth cranial nerve involvement. In addition, several (42.9%) patients complained of taste changes. Migraine is associated with MRS in various studies from 41 to 47%, however, in others the prevalence did not surpass 10-20%. [9] In our study 23.8% patients complained of headache and 14.3% of migraine, which is similar to the percentage reported (14%) by Greene et al. [10] Our findings suggest that the presence of migraine, pain, tinnitus, dizziness, and dysgeusia symptoms were unrelated to whether the patient had the full MRS triad.

The mean interval between episodes was 4.7 years (range 0-30, median 3). Six (28.5%) of the patients reported a family history of MRS.

The mean number of episodes of facial paralysis in our group of patients was 3.14, which is higher than the number reported in other studies. [9] However, it is difficult to compare our data to other studies as this number is dependent to the follow-up time of the patients. Excluding the 4 patients who only had 1 episode of facial paralysis, the mean time between episodes for the remaining 17 patients was 4.7 (range 0 to 30) years.

The highest number of episodes of facial paralysis per patient in our series was 8 over 29 years. The mean number of episodes of facial paralysis in our group of patients was 3.14, which is higher than the number reported in other studies. [9] However, it is difficult to compare our data to other studies as this number is dependent to the follow-up time of the patients. Excluding the 4 patients who only had 1 episode of facial paralysis, the mean time between episodes for the remaining 17 patients was 4.7 (range 0 to 30) years.

Methods: We performed a retrospective review of the patients diagnosed with MRS at a University based Facial Nerve Center.

Results: Twenty-one patients were identified from 1971 to 2010. The age of presentation ranged from 22 to 67 years (mean 44.1). Seven (33.3%) were male and 14 (66.7%) were female. All (100%) patients had facial paralysis. Fourteen (66.7%) patients who initially presented with unilateral paralysis subsequently developed metachronous contralateral paralysis (alternating unilateral facial paralysis). One (4.7%) patient had simultaneous bilateral facial paralysis. The number of episodes per patient ranged from 1 to 8 (mean 3.1). The patient with most episodes of facial paralysis had 4 on the left and 4 on the right (metachronous). The age at first incidence of facial paralysis ranged from 2 to 60 years (mean 34.4, median of 39). The mean interval between episodes was 4.7 years (range 0-30, median 3). Six (28.5%) of the patients reported a family history of MRS.

Conclusions: MRS is a rare disease of unknown pathogenesis in which oligosymptomatic forms predominate. Patients with this disease may present to differenct specialties complaining of different symptoms, and frequently, not all the classic features of the triad will be present. In our series of facial paralysis patients diagnosed with Melkersson-Rosenthal Syndrome, a higher proportion had the full triad of symptoms than has been previously reported in the literature.

METHODS AND MATERIALS

We searched for patients with MRS in the senior author’s database of the Facial Nerve Center of the Department of Otolaryngology, University of Pittsburgh.

RESULTS

Twenty-one patients were identified from 1971 to 2010. The age of presentation ranged from 22 to 67 years (mean 44.1). Seven (33.3%) were male and 14 (66.7%) were female. All (100%) patients had facial paralysis. Fourteen (66.7%) patients who initially presented with unilateral paralysis subsequently developed metachronous contralateral paralysis (alternating unilateral facial paralysis). One (4.7%) patient had simultaneous bilateral facial paralysis. This patient had 2 more episodes of unilateral facial paralysis (1 left, 1 right) that occurred subsequent to the diagnosis of MRS. The number of episodes per patient ranged from 1 to 8 (mean 3.1). Laterality was relatively equal: The patient with most episodes of facial paralysis had 4 on the left and 4 on the right (metachronous). This was followed by 3 patients with 6 episodes each. The age of first incidence of facial paralysis ranged from 2 to 60 years (mean 34.4, median of 39).

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REFERENCES

1. Visinescu, L.M. and D.A. Karmann. The Melkersson-Rosenthal syndrome. Facial Nerve Centre, University of Pittsburgh. Pittsburgh. Each patient’s data was reviewed, extracted, and de-identified in accordance with institutional protocols. The following variables were recorded and analyzed: age, sex, number of episodes of facial paralysis, laterality, House-Brackmann score, lingua plicata, facial edema, facial paresthesia, history of diabetes, migraine/headache, tinnitus, dizziness, dysgeusia, surgical treatment and family history.

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Melkersson-Rosenthal Syndrome is a rare disease of unknown pathogenesis in which oligosymptomatic forms predominate. Patients with this disease may present to different specialties complaining of different symptoms, and frequently, not all the classic features of the triad will be present. In our series of facial paralysis patients diagnosed with Melkersson-Rosenthal Syndrome, a higher proportion had the full triad of symptoms than has been previously reported in the literature.