

Melkersson–Rosenthal Syndrome with Isolated Immunoglobulin E Hypogammaglobulinaemia

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Melkersson–Rosenthal syndrome (MRS) is a rare disorder classically presenting with a triad of recurrent orofacial swelling, facial palsy and a fissured tongue. This case report describes a case of MRS in a patient with isolated immunoglobulin E (IgE) hypogammaglobulinaemia. The 52-year-old woman presented with puffy

eyelids, lower lip swelling and right facial nerve palsy. Fissures of the tongue were also noted. On investigation she was found to have a markedly low serum IgE level. This case report clearly indicates that IgE-mediated reactions do not play a significant role in the development of MRS.

KEY WORDS: MELKERSSON–ROSENTHAL SYNDROME; FACIAL PALSY; FISSURED TONGUE; OROFACIAL SWELLING; ISOLATED IMMUNOGLOBULIN E DEFICIENCY

Introduction

Melkersson–Rosenthal syndrome (MRS) is a rare disorder classically presenting with a triad of recurrent orofacial swelling, facial palsy and a fissured tongue;^{1,2} the complete triad is seen only infrequently. We describe here the first reported case of the triad of MRS symptoms in a patient with isolated immunoglobulin E (IgE) hypogammaglobulinaemia.

Case report

The case involves a 52-year-old woman who had been born to healthy parents at 40 weeks of gestation, with no complications of pregnancy or delivery. After an episode of encephalitis at the age of 3 months, her development was markedly delayed; she could sit alone and walk with support, but did not speak any meaningful words.

At the age of 50 years she suffered from an odontogenic infection which was successfully treated with antibiotics. At 51 years of age her care nurses reported that once a month she would lose her appetite and interest in her surroundings, and would lie recumbent in a seemingly exhausted condition for 3 – 4 days. One year after the appearance of these symptoms, the patient presented with puffy eyelids, lower lip swelling (Fig. 1) and right facial nerve palsy. Fissures of the tongue were also noted (Fig. 2). The orofacial oedema and facial nerve palsy showed a recurrent course and were accompanied by the apparent exhaustion. The patient was post-menopausal.

A biopsy specimen from the lower lip showed mild perivascular inflammation but no epithelioid granulomas or multinucleated giant cells. Liver and kidney



FIGURE 1: Swelling of the lower lip in a patient with Melkersson–Rosenthal syndrome



FIGURE 2: Fissured tongue in a patient with Melkersson–Rosenthal syndrome

function tests as well as a complete blood count were normal. Immunoglobulin and complement levels are given in Table 1. The serum IgE value was markedly low and remained low on consecutive examinations. The IgA and 50% haemolytic unit of complement values were above the normal range. Serum angiotensin-converting enzyme activity and chest X-ray were normal.

Based on the presence of unilateral facial nerve palsy, recurrent orofacial oedema and fissured tongue, a diagnosis of MRS was made. The non-specific inflammation that was seen in our patient's lower lip was consistent with MRS, and excluded possible underlying conditions such as Crohn's disease, sarcoidosis, amyloidosis, and other granulomatous or degenerative disorders. MRS may be associated with various complications, including trigeminal neuralgia, other cranial nerve symptoms and migraine. We speculated that our patient's apparent exhausted condition may have been caused by migraine, but this could not be confirmed or ruled out given her communication disorder.

Discussion

No specific test is diagnostic for MRS. Zimmer *et al.*³ reported on 28 biopsy specimens from 26 patients: 46% showed

granulomatous inflammation, 36% showed non-specific inflammation, 11% showed only incidental findings and no abnormalities were seen in 7%.

The aetiology of MRS is unknown. Reported associations include Crohn's disease, allergy, angio-oedema, odontogenic infection and menstrual conditions.² The clinical significance of the known protective role of IgE in host defence mechanisms remains unclear. Schoettler *et al.*⁴ described a family with sinopulmonary disease and isolated IgE hypogammaglobulinaemia. Of the 14 symptomatic family members, 12 showed a markedly low IgE concentration (< 5 IU/ml), whereas all the asymptomatic individuals had a normal IgE concentration (> 10 IU/ml). In contrast, Levy and Chen⁵ reported an IgE-deficient person who was healthy. The patient in the present case report did not have manifestations similar to those seen by Schoettler *et al.*⁴

This case report clearly indicates that IgE-mediated reactions, which include some specific types of allergy, do not play a significant role in the development of MRS. Further investigation of IgE concentrations may help reveal the pathological mechanisms of MRS, although the possibility that the association between MRS and isolated IgE hypogammaglobulinaemia in

TABLE 1:
Immunoglobulin (Ig) and complement (C) levels in a patient with Melkersson–Rosenthal syndrome

Measure	Observed	Normal
IgE (IU/ml)	3.45	>5
IgG (IU/ml)	1379	680 – 1650
IgA (IU/ml)	494	84 – 438
IgM (IU/ml)	183	57 – 288
C3 (mg/dl)	112	86 – 160
C4 (mg/dl)	28	17 – 45
50% haemolytic unit of C in sera (U/ml)	55.0	25.0 – 48.0
C1q (µg/ml)	< 1.5	< 3.0

our patient occurred by chance cannot be excluded given that an elevated IgE concentration has also been reported in MRS.¹

Conflicts of interest

No conflicts of interest were declared in relation to this article.

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