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Anesthetic management of patients with Melkersson Rosenthal syndrome

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Abstract
Melkersson Rosenthal Syndrome (MRS) is a rare disorder characterized by relapsing facial paralysis, persistent or recurrent orofacial edema, and lingua plicata. It may cause difficult airway, drug allergy, and angioedema. In our anesthetic management of two patients with MRS, preanesthetic immunological blood examination and skin tests for hypersensitivity to anesthetic drugs were applied. Because the principal goal is to avoid all factors that may stimulate, an allergic reaction, anesthetic drugs known to trigger urticaria were avoided. Body and operating room temperatures, changes of which may trigger allergic reactions, were kept constant during the perioperative period. Emergency precautions were taken for probable angioedema. MRS is a rare syndrome, and if its manifestations are misunderstood as simple facial paralysis, it may be overlooked by anesthesiologists. Anesthesiologists must be careful of several problems in patients with MRS.

Key words Melkersson Rosenthal syndrome · Anesthesia

Introduction

Melkersson Rosenthal syndrome (MRS) is a rare granulomatous inflammation, of unknown etiology, characterized by relapsing facial paralysis, persistent or recurrent orofacial edema, and lingua plicata [1–5]. MRS occurs more often in females than in males, with a ratio of nearly 3:1 [6]. Typically, paralysis occurs transiently at the site involved and is indistinguishable from Bell's palsy [7,8]. The duration is short at first, subsiding completely in a few hours or days, therefore simulating angioedema [8].

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Although there have been previously published reports about MRS in the literature, we could not find a report about the anesthetic management of patients with this syndrome. Here, we report two patients with MRS and discuss the anesthetic management of patients with MRS.

Case 1

A buccal cheiloplasty was planned for a 31-year-old man (175 cm, 83 kg, American Society of Anesthesiologists [ASA] I) who was diagnosed with MRS (Fig. 1). Swelling of lips and cheek, palpable edema with local tenderness in the right preauricular region, nodules in the cheeks and nasal bridge, fissure of the tongue, and right-sided facial paralysis were revealed by preoperative physical examination.

Case 2

A 16-year-old girl (158 cm, 46 kg, ASA I) had suffered from a permanent hemifacial swelling on the right side for 4 years. She had neither lingua plicata nor evidence of facial palsy, although she had had an attack of right facial paralysis when she was 13 years old. Suction lipectomy to correct the facial asymmetry and to reduce the buccal swelling was planned, to be performed with the patient under general anesthesia.

Procedure

Both of the patients' baseline laboratory investigations were normal, including complete blood count and serum biochemistry. C3, C4, IgE1, IgG, IgM, and total IgE levels were in the normal ranges. No abnormal findings were seen on head and neck magnetic resonance imaging

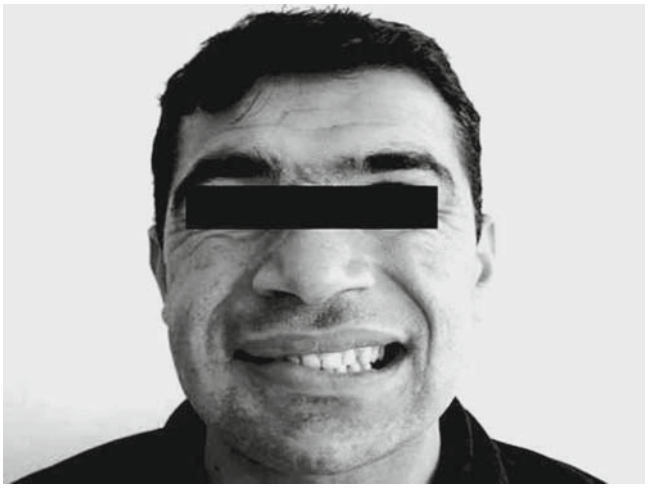

Fig. 1. A 31-year-old man with Melkersson Rosenthal syndrome

(MRI) and electromyography (EMG). Intradermal skin tests for hypersensitivity to anesthetic drugs (etomidate, propofol, fentanyl, cisatracurium, vecuronium) and prick tests to other allergens (milk, egg, house dust mite, grass, hazelnut, peanut, mixed herbal products, etc.) were negative (0.1 ml of induction agent diluted to 1:100; muscle relaxant diluted to 1:1000, opioid diluted to 1:100000).

Necessary precautions against potential allergic reactions were taken before the induction of anesthesia. Methylprednisolone 1 $mg \cdot kg^{-1}$ was given to each patient just before induction. Adrenaline-soaked gauze was applied to the nasal cavity to prevent bleeding. The induction of anesthesia was performed with fractional doses. In total, 0.3 $mg \cdot kg^{-1}$ etomidate, 2 $\mu g \cdot kg^{-1}$ fentanyl, and 0.1 $mg \cdot kg^{-1}$ cisatracurium were given. A fiberoptic bronchoscope, was prepared but we did not need it. The patients were easily intubated with a cuffed tracheal tube (7.5-mm internal diameter for case 1, and 6.5 mm for case 2), in a nasotracheal manner. No nasal bleeding was observed. Anesthesia was maintained with isoflurane 1% in a mixture of 40% oxygen and 60% nitrous oxide. The body temperatures of the patients (36.5°C–37°C), and operating room temperatures (24°C–25°C) were kept constant because changes of temperature could trigger urticaria. Warm blankets were used for the patients during the perioperative period. Hemodynamic measurements were stable during the operation. At the end of the operation, the patients were awakened and extubated uneventfully. Both patients were observed for 60 min in the recovery room.

Discussion

MRS is a rare disease with oral swelling, mostly presenting in the first two decades of life, and associated with facial nerve paralysis and lingua plicata. Its etiology consists of infectious agents, allergic reactions to foods and supplements, and immunological and genetic factors [2–5]. Allergic reactions may appear as urticaria and angioedema. Angioedema is an urticarial form that is associated with edema of the lips or mucosa of internal organs, or it may affect layers of the skin. The nasopharyngeal mucosa may be affected and swelling of the tongue and further laryngeal edema is possible. Patients may die due to cardiogenic shock and hypotension [9,10].

A diagnosis of urticaria and angioedema has often been made before the anesthesiologist contacts the patient. If any systemic disorders are responsible for the skin lesions, then a careful history should be taken and physical examination for disease associated with angioneurotic edema should be carried out. Previous medical records must be obtained to examine the course of any previous anesthetic exposure [9].

The incidence of hypersensitivity in anesthetic practice varies from about 1:10000 to 1:20000 [11]. Of note, barbiturates may cause direct histamine release. Propofol has caused anaphylaxis. Hypersensitivity to benzodiazepines and etomidate is rare, and these drugs do not cause direct histamine release. Muscle relaxants are the most common cause of hypersensitivity in anesthetic practice. Succinylcholine is the most immunogenic, although reactions to vecuronium, atracurium, alcuronium, gallamine, and d-tubocurarine are well documented [11]. Cisatracurium does not cause histamine release [12]. In addition, anaphylaxis has been reported with most opioids. Fentanyl is a rare cause of anaphylaxis and does not lead to significant histamine release. Hypersensitivity to local anesthetics is also rare [11].

The principal goal in the management of patients with urticaria and angioedema under anesthesia is to avoid all triggers of these manifestations. Drugs that are known to trigger urticaria should be avoided. If temperature is the trigger of urticaria, then the operating room should be warmed before the patient arrives, and all facilities for maintaining temperature control should be made available. In addition, mucosal lesions in the nasopharyngeal airway should be examined before intubation [9].

For these reasons, with the consideration of the presence of an allergic background, we applied immunological and skin tests in our patients. The body temperatures of the patients and operating room temperatures were kept constant, because changes of temperature can trigger urticaria. Etomidate, fentanyl, and cisatracurium

were administered in the induction of anesthesia, because of the known safety of these drugs. Maintenance of anesthesia was safely provided by taking emergency preventive measures for probable angioedema.

In conclusion, MRS is a rare syndrome, and if its manifestations are thought to be simple facial paralysis, and not investigated enough, it may be overlooked by anesthesiologists. Anesthesiologists must be careful and consider the probability of several problems such as difficult airway, drug allergy, and angioedema. However, a safe and successful method of anesthetic management for the patient with MRS has yet to be established. Further case reports are needed.

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