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Anesthetic management of a child with olmsted's syndrome

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Introduction

In 1927, Olmsted described a 5-year-old boy in whom palmoplantar keratoderma had developed during the second half of his first year of life, and who later presented with circumnasal, circumoral, and circumanal hyperkeratotic plaques [1]. Since then, only three cases have been reported in the English literature [1–3].

We experienced anesthetic management of a child with Olmsted's syndrome undergoing excision of hyperkeratotic plaque. We review the literature describing Olmsted's syndrome and discuss potential anesthetic problems of patients with this rare syndrome.

Case report

A 12-year-old boy (height 115 cm and body weight 19 kg) was admitted to our hospital for treatment of palmar hyperkeratosis. He was normal at birth. Hyperkeratotic plaques developed around the lower lip at 2 weeks of age, and on the fingers and toes at 6 months of age, gradually extending onto the palms and soles. Prior examination revealed painful thick hyperkeratotic plaques resembling a cast of the hands and linear extension onto the flexor surfaces of the wrists; flexion deformities of the fingers; leukokeratosis of the tongue; alopecia on the left forehead; and hyperflexibility of the joins. Roentogenograms showed osteolysis of the distal phalanges of the fingers and toes and maldevelopment of the mandibula and maxilla. His bone age (which was determined about one year before he underwent surgery) was 8 years at a chronological age of 11 years. His mental development was average for his age, and neurological and ophthalmologic examinations revealed no abnormality. Oral etretinate produced reduction of pain of the hands and feet with mild improvement of the skin lesions, but it was discontinued due to perianal erosion and cheilitis. The patient underwent excision of the hyperkeratotic plaque and skin graft under general anesthesia at ages 4, 11, and 12. At 4 years of age (height 93.5 cm and body weight 13 kg), he was scheduled for excision of the left sole. He was premedicated with diazepam 6 mg and atropine 0.25 mg p.o., subsequently anesthetized with halothane and nitrous oxide (N_2O) in oxygen via a face mask without complications. Venous access was easily established at the forearm.

At 11 years of age (height 112 cm and body weight 18 kg), hyperkeratotic plaques of palms and soles were removed. Following premedication with diazepam 7.2 mg and atropine 0.5 mg p.o., anesthesia was induced with halothane and N_2O in oxygen via a face mask. Muscle relaxation was obtained with 2 mg of vecuronium. His trachea was intubated with nasotracheal tube without difficulty. Anesthesia was maintained with halothane and N_2O in oxygen.

At 12 years of age (height 115 cm and body weight 19 kg), hyperkeratosis of the left palm was excised with skin grafting. Premedication consisted of diazepam 7.6 mg and atropine 0.55 mg p.o. Anesthesia was induced with sevoflurane and N_2O in oxygen via a face mask. Intravenous 2 mg of vecuronium facilitated orotracheal intubation, and the endotracheal tube was secured with adhesive tape above the upper lip and around the mandible and cheek to keep the hyperkeratotic plaque of the lower lip from coming into contact with the tape. Anesthesia was maintained with sevoflurane and N_2O in oxygen.

Our careful management on laryngoscopy and insertion of the thermometer probe into the rectum pre-

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vented development of perioral or perianal fissure. All postoperative courses were uneventful.

Discussion

In Olmsted's syndrome, hyperkeratotic changes of the palms, soles, and around all of the body orfices occur. It is a very rare disease with only three cases reported in the English literature [1–3]. These patients received many treatments: topical medications and various oral agents, including etretinate, vitamin A, antihistamines, biotin, vitamin E, iodoquinol, and antimicrobial therapy. However, these therapies were not significantly effective, except for oral or topical etretinate, both of which were discontinued in several cases due to adverse effects. Thus, the most reliable therapy may be shaving the thickened epidermis, but this causes discomfort. Severe hyperkeratosis possibly causes contracture of fingers and ankles (flexion deformity of the fingers or equine foot), osteolysis of phalanges, or spontaneous amputation of the fingers or toes. Thus, excision of the plaque and skin graft under general anesthesia has been employed.

Table 1 summarizes the three cases of Olmsted's syndrome described in the English literature [1–3]. Characteristic symptoms and signs of the syndrome are: (1) painful hyperkeratotic plaque of palms, soles, and periorifices, (2) dystrophic nail, (3) contracture deformity or osteolysis of fingers and toes, (4) leukokeratosis of the tongue, (5) hair loss, (6)hyperflexibility of the joints, (7) high-frequency loss of hearing, and (8) retardation of bone age. Though the small number of reported cases does not allow us to regard each of these as an essential manifestation for diagnosis, findings (2) and (3), in addition to (1), may be the principal diagnostic criteria, since they were observed in all cases.

Patients with Olmsted's syndrome need repeated anesthetic management from their early childhood. Frequent hospitalizations and medical interventions caused our patient to become agitated during anesthetic induction. If we had provided more empathetic support, induction of anesthesia with less psychological trauma might have been possible for our patient. Severe perioral and perinasal hyperkeratotic plaque may cause difficulties in securing the endotracheal tube. In our case, as perioral hyperkeratosis was restricted to a small area under the lower lip; we successfully fixed the endotracheal tube by taping the tube to the face while avoiding the plaque. Patients with a wide range of perioral hyperkeratosis need special care in securing the endotracheal tube; nasotracheal intubation or fixation methods without using tapes or adhesives may be needed. Rough laryngoscopy may cause perioral fissures. Careful management of patients is fundamental and should be emphasized in patients with Olmsted's syndrome, who may easily suffer from fissures of hyperkeratotic plaques by inattentive handling. Although this caution may seem minor, it is nonetheless an important factor in improving the quality of anesthetic management. No systemic complications have been known as principal manifestations of Olmsted's syndrome. In the German literature [4], primary sclerosing cholangitis and immunodeficiency were reported

Table 1. Clinical manifestations of Olmsted's syndrome in the English literature

Authors (year of publication)	Olmsted (1927)	Poulin (1984)	Atherton (1990)	Nishina Present study
Age (years)	5	?	4	12
Age of onset	2.6 years	at birth	1.4 years	2 weeks
Short stature	+	+	+	+
Delayed social age	+	NI	+	−
Delayed bone age	NI	NI	+	+
Hyperflexibility of joints	NI	+	NI	+
Alopecia	−	+	NI	+
Auditory disturbance	NI	+	NI	−
Leukokeratosis of the tougue	NI	+	NI	+
Hyperkeratosis				
Palms and soles	+	+	+	+
Perinasal	−	+	+	+
Perioral	+	+	+	+
Perianal	+	+	+	+
Contracture of fingers	+	+	+	+
Dystrophic nail	+	+	NI	+
Osetolysis of the phalanx	NI	+	NI	+

NI, not indicated

in a patient with Olmsted's syndrome. However, the relationship between these disorders and Olmsted's syndrome remains unclear.

In conclusion, this syndrome has not been reported to be accompanied by systemic complications, which may affect anesthetic management. However, it is our belief that careful handling of a patient with Olmsted's syndrome during perioperative periods is important to avoid skin injuries such as periorificial fissure.

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