

A Case of Kawasaki Disease With Severe Lip and Oral Mucosa Involvement Complicated With Microstomia and Corrected With Surgery

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Kawasaki disease (KD) is an acute self-limited systemic vasculitis that affects medium-sized arteries.¹ Diagnosis is based on clinical features including fever for more than five days accompanied by bilateral conjunctivitis, oropharyngeal changes, cervical lymphadenopathy, polymorphous rash, and peripheral extremity changes.^{1,2} Conjunctivitis, oropharyngeal changes, and rash are the most common clinical features of the disease.³ Changes of the lips and oral cavity include erythema, dryness, fissuring, peeling, vertical cracking and bleeding of the lips; a strawberry tongue with erythema and prominent fungiform papillae; and diffuse erythema of the oropharyngeal mucosa.² In nearly all of the cases, these findings of the lip and oral mucosa heal without any sequela. In this article, we present a case of KD with extensive and severe lip and oral mucosa involvement complicated with microstomia that needed surgical correction.

A six-year-old boy was admitted to the hospital because of fever and myalgia for six days and rash for three days. On physical examination, he had bilateral nonpurulent conjunctivitis, perineal erythema, erythematous macular rash on the

trunk and extremities, edema of the palms and soles, red and cracked lips with diffuse pharyngeal hyperemia without any exudate. On laboratory tests, leukocyte count was 17,300/mm³, platelet count was 550,000/mm³, C-reactive protein (CRP) was 235 mg/L (normal 0-5 mg/L), and erythrocyte sedimentation rate (ESR) was 88 mm/hour. The child was diagnosed as having KD and intravenous immunoglobulin (IVIG) 2 g/kg was given and salicylate (80 mg/kg/day) was started. Initial echocardiographic examination did not show any coronary artery abnormality. As fever did not subside after 48 hours of IVIG infusion, a second dose of IVIG (2 g/kg) was given. The patient became subfebrile after the second dose of IVIG but developed extensive lip and oral mucosa involvement with dense hemorrhagic crusts on the lips. As the fever did not resolve completely on the eighth day of hospitalization, the child was referred to our institution with the diagnosis of IVIG-resistant KD. He had minimal perineal desquamation, and swollen, blackish, crusted, bleeding lips (Figure 1a). Although the mouth involvement resembled Steven-Johnson syndrome/toxic epidermal necrolysis like picture,

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Figure 1. (a) Swollen, crusted, bleeding lips on second week, (b) development of microstomia on second month, (c) view of lips two years after surgery.

the remaining skin of the body did not have any erythema multiforme like rash, vesicles, bullae, or sloughing. As he still had fever, thrombocytosis (platelet $880,000/\text{mm}^3$) and high acute phase reactants (CRP: 120 mg/L, ESR: 90 mm/hour), three days of high-dose methylprednisolone (30 mg/kg/day) followed by 2 mg/kg/day were started. Viral and bacterial serologic studies did not reveal any positive result. Also blood, urine, and swab cultures from the mucosa of the lips and mouth did not yield any microorganism. He did not develop any symptom, sign or laboratory abnormality of the upper and lower respiratory tract, lung or kidney involvement. Control echocardiographic examination showed hyperechogenicity on the left coronary artery wall without ectasia. On the seventh day of prednisolone treatment, the fever subsided. Prednisolone treatment was discontinued on the third week with gradual tapering. Oral mucosa and lip involvement healed gradually in one month and echocardiographic examination was normal by all parameters. The child was discharged with salicylate (3 mg/kg/day) treatment that was given for two months. On the second month of the follow-up, he was unable to open his mouth fully and it was seen that he developed microstomia (Figure 1b). He had reconstructive lip surgery with bilateral commissuroplasty and autologous fat injection into the lower lip and vermillion border by the plastic and reconstructive surgery team.

Microstomia defines a condition of reduction in the size of oral aperture which can be either acquired or congenital.⁴ The most common cause of acquired microstomia in healthy children seems to be electrical, thermal, or chemical burns.⁵

There are some rare genetic diseases that have a component of microstomia like Freeman–Sheldon syndrome, otocephaly, and dystrophic epidermolysis bullosa.⁵ The most well-known rheumatic disease associated with microstomia is systemic sclerosis.⁶ Tuncer et al.⁷ presented a two-and-a-half-year-old boy with severe microstomia due to leukocytoclastic vasculitis of unknown origin. Apart from microstomia, the case of Tuncer et al.⁷ also demonstrated gangrene with autoamputation in toes and fingers. There are a few reports of KD patients with peripheral gangrene and autoamputation.⁸

Intravenous immunoglobulin (2 g/kg, 12 hours of infusion) is the standard treatment modality in KD.^{1,2} If fever does not resolve or it recurs after 36–48 hours of the first IVIG infusion, the patient is called as IVIG-resistant. There is no consensus on the treatment of such patients. A trial of second dose IVIG (2 g/kg) is recommended.^{1,3} Corticosteroids may also be an alternative in IVIG-resistant cases. Starting with high dose methylprednisolone pulse (30 mg/kg/day) therapy for one–three days, continuation with 2 mg/kg/day, and discontinuation in two–three weeks with gradual tapering are recommended.^{9,10} Our case had already failed two infusions of IVIG and we successfully treated him with corticosteroids. Therefore, we think that corticosteroids may be a good and easily accessible option in the treatment of IVIG-resistant KD patients.

The child is being followed-up for two years and he did not develop any sign of other underlying chronic vasculitis or systemic sclerosis and has near normal mouth opening (Figure 1c). To the

best of our knowledge, this is the first case of KD with extensive and severe lip and oral mucosa involvement complicated with microstomia.

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