

KARAPANDZIC FLAP FOR RECONSTRUCTION OF LOWER LIP IN A 18-MONTH OLD BOY WITH CONGENITAL AGAMMAGLOBULINEMIA AND ECTHYMA GANGRENOSUM. CASE REPORT

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Abstract. *The reconstruction of lower lip in children is extremely rare and challenging procedure. The etiology in literature reveals trauma and infection. An 18-month boy was admitted with sepsis and pneumonia. Laboratory tests revealed congenital agammaglobulinemia. Necrosis of lower lip developed and was diagnosed as ecthyma gangrenosum. Blood culture was positive for Pseudomonas aeruginosa. Multiple abscess formations were found in abdominal wall and gluteal region and were treated by incisions. After spontaneous demarcation of necrotic tissue in lower lip the Karapandzic flap technique was used for reconstruction. Karapandzic flap can be used as optimal method for reconstruction of lower lip in children with satisfactory functional and aesthetic results.*

Key words: *Karapandzic flap, ecthyma gangrenosum, reconstruction*

Introduction

The congenital agammaglobulinemia is inherited disease characterized by depletion of humoral immune response. The diagnosis is confirmed by low concentrations of blood immunoglobulins (IgG or IgM). In any case of immunocompromised pediatric patient, including aplastic anemia, leukemia or agammaglobulinemia, the high susceptibility for infections is recognized, mainly in the first four years of life. The key treatment is intravenous administration of immunoglobulins which boosts immune system [1].

One of the most common skin infections in immunocompromised children is ecthyma gangrenosum (EG). The EG can be a manifestation of any immunocompromised disease, such as hematological malignancies, severe burns, chemotherapy or immunodeficiency syndromes, including congenital agammaglobulinemia. Ecthyma gangrenosum is commonly associated with Pseudomonas aeruginosa bacteremia. It is almost always a sign of Pseudomonas sepsis. The clinical manifestation is recognized as hemorrhagic pustules or infarcted skin appearing area with surrounding erythema. EG was first described in 1897 by Barker. Histology reveals necrotizing vasculitis. The final appearance is necrotic skin plaque surrounded by erythema [2, 3]. The lesion is mostly located in gluteal and perineal regions (57%), extremities 30%, trunk 6% and face 6% [3, 4]. The

antibiotic therapy is the first treatment for initial lesion and must be defined according to blood culture sensitivity. Necrotic lesions must be treated by surgical debridement [5]. The most challenging are the locations of EG in the facial area where esthetic and functional requirements must be fulfilled [5, 6].

In case of lower lip defect, different reconstructive options must be taken into consideration. The goals are both aesthetic result and functional competence of lower lip (speech and food intake). The lip reconstruction can be performed using local flaps or microvascular free tissue transfer. One of the local flaps was described by Karapandzic and this flap was defined as myoneurovascular pedicled advancement flap [7]. The full-thickness lip defect requires reconstruction of all layers (mucosa, muscle, skin) and every case should be approached separately. The tissue from the cheek and the rest of the lower lip used for Karapandzic reconstruction most closely resembles the missing part of lower lip according to texture and color. Musculocutaneous flaps from both sides of the defect have sufficient height to restore the height of the lower lip and motor nerves are spared for appropriate functioning of orbicularis oris muscle. This flap is good for single stage lower lip reconstruction but it has significant limitations in microstomia [8].

Our aim was to present the young patient with ecthyma gangrenosum treated with Karapandzic flap technique.

Case Report

An 18-month boy was admitted in pediatric clinic with sepsis and bilateral pneumonia. Laboratory tests revealed

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congenital agammaglobulinemia. During the hospital stay, he developed multiple subcutaneous abscesses in the abdominal and gluteal region, as well as an abscess and necrotizing lesion in the central part of the lower lip diagnosed as ecthyma gangrenosum.

Blood culture was positive for *Pseudomonas aeruginosa* and the antibiotics were administered according to blood culture sensitivity (Ceftazidime and Gentamycin). The immunoglobulins were administered. After 10 days, the patient's general health condition was stable. Initial surgical treatment included multiple incisions of abscesses and removal of necrosis from left groin region. The vasculitis in lower lip lead to necrosis of the central part of the lower lip. After spontaneous demarcation, initial scarring occurred (Fig. 1).

The defect in lower lip was reconstructed under general anesthesia using the Karapandzic flap technique. The postoperative result is shown in Figure 2.

Discussion

Ecthyma gangrenosum is a skin infection caused mostly by *Pseudomonas* and manifested as skin necrosis surrounded by erythema [9–11]. It occurs in 1% to 30% of cases of *Pseudomonas* sepsis [12]. Other described etiology includes *Klebsiella oxytoca* or *Escherichia coli* and in this case an early broad spectrum antibiotic regimen is recommended for initial treatment of ecthyma gangrenosum [13, 14]. Lesions can occur anywhere but are more common in the perineum, buttocks, axillae, and extremities, but in our case they appeared in the central part of lower lip [15]. In some cases of ecthyma gangrenosum even facial nerve palsy with lesion in external auditory canal developed [16]. Surgical repair was planned when the lesion became stable [15].

In order to avoid excision of healthy tissue, the lower lip was not treated before spontaneous demarcation. The reconstruction of full thickness of the lower lip is a challenging procedure especially in children. The ecthyma gangrenosum occurs in the facial region in only 6 % of all cases. Spontaneous demarcation occurred after two weeks, leaving a full-thickness defect in the central part of the lower lip. Why did we perform the Karapandzic technique? In our opinion, it is the most acceptable technique for children because the myocutaneous flaps had the same skin color and texture as the missing part. The other reason is preservation of motor nerves. The operative procedure takes short time and surgery fulfills aesthetic requirements.

Some authors support the use of the Karapandzic flap, in its original form or modified, but some prefer labiomental flap, double rectangular rotation flap, the

Fujimori gate flap from nasolabial area or staircase (step) technique [17–24]. Despite the fact that poor prognosis is associated with multiple lesions, as well as delayed treatment and neutropenia, our case had acceptable recovery [25]. Any reconstruction of the lips must include both functional and cosmetic considerations [19].



Fig. 1. Preoperative central lip defect



Fig. 2. Postoperative result after Karapandzic flap technique.

Conclusion

Satisfactory esthetic and functional results were obtained in a child with congenital agammaglobulinemia and ecthyma gangrenosum using Karapandzic flap technique for central part of the lower lip.

In our knowledge, this is the youngest patient treated with the Karapandzic flap technique.

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