CASE REPORT

Management of Rare Facial Cleft Tessier-7 : A Case Report

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ABSTRACT

Macrosomia or transverse facial, which is usual we called as Tessier 7 congenital cleft or lateral facial cleft is a birth defect characterized by the patient's mouth being wider than the average normal size. Macrosomia can be part of a syndrome, such as Goldenhar syndrome, Treacher Collin syndrome or Hemifacial Microsomia. This condition happened due to bad fusion of the mandible and maxillary bone during embryonic development. This disorder dominated by male, often unilateral form, rarely bilateral. We report a 10-month-old male patient with unilateral cleft in the left corner of the lip. The patient was born of 32-year-old mother with a history of P3A0 pregnancy. The patient was born with a normal delivery process assisted by an obstetrician at General Hospital at Tasikmalaya area. The baby was born at the gestational age of 38 weeks and weight of 2,6 kg. The patient has no family history of cleft lip and palate. We diagnosed this patient with Unilateral Tessier 7 Facial Cleft. In common, this case was treated with Z-plasty or other kind of methods, although in the end not expected scar may result. This time, we present a review of literature and a case of unilateral Tessier 7 cleft repaired with Z-plasty technique to minimize the linear scar and to expand the surgical preferences.

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INTRODUCTION

Macrosomia or transverse facial cleft is a unique type of craniofacial microsomia. It is a part of Tessier 7 facial cleft and happened in about 1:60,000 to 1:300,000 live of births. It is a result of abnormal fusion of the first and second branchial curves (1). Macrosomia can be accompanied with another craniofacial abnormalities. It also can be a part of syndromes such as Treacher Collins and Goldenhar syndrome/Oculo-Auriculo-Vertebral Spectrum (OAVS). Transverse facial cleft can be either one sided or two sided. Severe macrosomia is a cleft which expand from the corner of the mouth to the ear and can involve the temporal and cheekbone (2). Most cases are unilateral macrosomia and only just 10% to 20% of cases are bilateral. Right side of commissure is more happens than the left, and men are influenced more happens than women (3). Butho and Botha recommend a four-lining closure technique with a primely for skin closure. It is important to know the right position of the cutaneous z-plasty to gain good facial expressions during smiling (4). The purpose of surgery is to improve a better articulation, eating process, aesthetic face, and concomintant psychological (5).

CASE REPORT

We reported a case of 10-month-old baby boy with complaint of large left side mouth and drooling saliva. The baby was born from a 32 years old mother with history of pregnancy P3A0 with a normal delivery process and gestational age of 38 weeks, helped by a midwife in Clinic at Sukabumi area with birth weight of 2,6 kg. There was no history of fall in pregnancy. There was history in consuming paracetamol and antibiotics in pregnancy. There was no history of cleft lip/palate in family. The patient has allergic in cold weather and has no history in surgery before. We also evaluate preauricular ear tag, on the other side there was no anomaly in other parameters such as occlusion and temporomandibular joint. There was no skeletal abnormalities in systemic evaluation. Hematology examinations are within normal limits. In repairing the ill-formed commissure, the baby was undergo the surgery under general anesthesia. There was no symmetrical face, gap at bilateral commisure sinistra with 3x1x0.5cm in size, abnormal shape of ear and low set ear, vesicular breath sound within normal limit and there was no rhonchi and wheezing (Figure 1).

The patient sign an informed consent agreement for treatment measures and approval to publish in scientific activities with ethical approval number: 2454/UN6.F.1/PT.00/2024. We treated this patient with Z-plasty technique. By the first, we marked the normal

commissure, then we positioning the midpoint of the upper lip to the middle of the peaks of the cupids bow. Midline of the upper lip, lower lip and columella of the nose should be in one straight line. On the upper lip, point A was marked and on the lower lip at the vermillion cutaneous junction, point B was marked. From the points A and B, we marked points X and Y 2mm from lateral. Incision were extended from the vermillion mucocutaneous junction to points X and Y (Figure 2).

From vermillion mucocutaneous junction of points X and Y, the mucosal triangular flaps were made by 45o oblique incision lines. Incision were made by following the marking and dissection were start from labial and mucosal sides to release the orbicularis oris muscle. In some cases, dog ear and V-shaped defect was formed because there was overlapped between muscle fibers

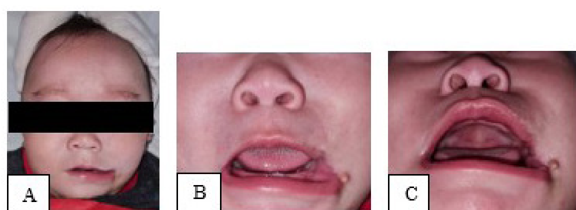

Figure 1: Preoperative photograph with left side ill-formed commissure with 3x1x0,5 cm in size. (A) Facial profil. (B, C) extra oral condition

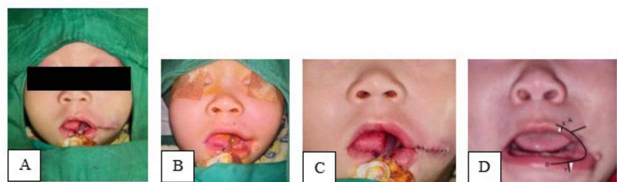

Figure 2: Intraoperative photograph with proposed incisions markings by modified Z-plasty technique. (A) Facial profil. (B,C) Final extra oral condition. (D) Design of Z plasty limbs and mucosal flaps

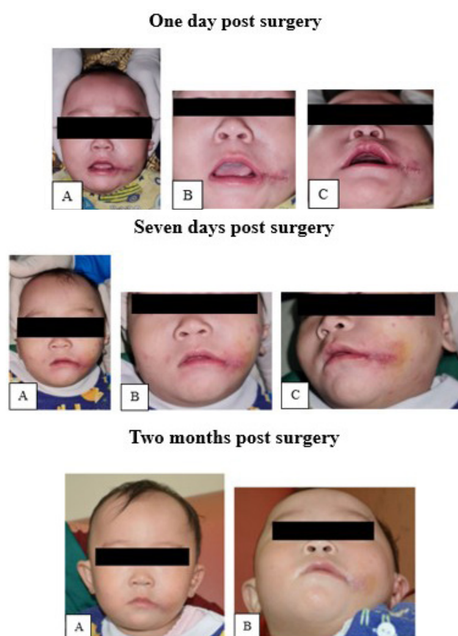

Figure 3: Postoperative photograph improvement condition day by day with commissure closest to symmetrical, minimal scar and natural look

at commissure of the upper and lower lip. One day postsurgery the commissure was closer to symmetrical. Seven days post surgery, photographs reveal that the commissure was reach to symmetrical state especially when the patient was smiling or even in rest condition. Two months post surgery, the clinical picture shows us good appearance with symmetrical commissure (Figure 3).

DISCUSSION

Macrosomia is a rare abnormalities with various kinds of clinical appearance. Deformities might classified from mild to severe. The Tessier 7 cleft are most often seen in combination with the OAVS involving the Goldenhar syndrome, Treacher Collins syndrome and Hemifacial microsomia (2). Orbicularis oris, buccinator and masseter muscle are involved in macrosomia's development. In surgical correction, there were many kinds of techniques described in the literature. Superiorly and inferiorly rotated, middle positioned, and agenic lateral are classified for Tessier 7 clefts by Butow and Botha in 2010. We have to consider the best technique to minimize the scar and blend it to natural skin wrinkles (4).

The best result of surgery is appropriate positioning of scars between different aesthetic region of face. Mild deformity shows clearly in this current case. According to achieve symmetrical face, reference points are marked from the abnormal side and into the normal side. There were many surgical techniques that have been offered in reconstructing macrosomia deformities. First, The Triangular skin flaps will give unnatural look and transpose skin at the commissure. Then The Vermillion square flap method might result poor scar at vermillion cutaneous junction of the commissure. Next, The Simple straight line closure with improper approximation of muscle bundles might result in fish mouth defect. Others, The W-plasty sometimes need undergo second revision surgery for more accurate result of the oral commissure. Z-plasty is more simple and has a comparable scar as W-plasty. Z-plasty become the best technique because it will minimize the defect and giving good wound closure (3).

Interestingly, aesthetic results and good functional were found as the outcomes of straight line closure combination with vermillion-mucosal flaps. Beside that, some writers polemize that the size of z-plasty would influence the final result. The more size of z-plasties (>10mm), the more scars will be seen. Apart from commissuroplasty, there were been many variations in the literature has been suggested. Rectangular and triangular vermillion-mucosal flaps are the most often used technique as in the literature. For intraoral mucosa closure, intraoral z-plasty or a straight line closure remains the best option. Z-plasty also helps in lengthening a webbed or camouflage scar and alter the position of a replaced anatomical point by

lifting or pushing it. In consideration of that each cleft is unique according to its anatomy (width, direction and length), there is no defined method for each step of the surgical procedure. To avoid unsatisfactory aesthetic and functionally outcomes, surgical details are important to known before surgery. Delayed healing and chronic ulceration are some of the complications by the transposed tissue from the upper to the lower lip. Z-plasty is more common used because of the delight and consistent result. According to Pan Zhou et al, twenty-one out of twenty-seven (78%) patients with hemifacial microsomia reach a prominent improvement (5).

CONCLUSION

In reconstructing macrosomia defects, there were many surgical techniques that have been offered. Z-plasty was used in this case because it is more simple and can camouflages scar. Afterall, the rarity of Tessier 7 cleft allows different approaches to be implemented and improved in order to achieve an optimal surgical result. Through this case, we try to expand surgical preferences in treating Tessier 7 clefts. Long-term follow-up could gives us different result from different closure methods and it would be a worth future studies.

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