

Lip-Nose Reconstruction Post-Cancrum Oris Defect: A Case Report

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Abstract

Cancrumoris is a disease of great antiquity described by Hippocrates and Galen. The term noma, a common synonym for the disease, is derived from the Greek verb 'to devour'. Tourdes (1948) defined cancrumoris that is still substantially correct. He described the condition as a gangrenous affection of the mouth that especially attacks children in whom the constitution is altered by bad hygiene and serious illnesses such as exanthematous fevers. According to him, the disease is manifested by an ulcer of the oral mucous membrane, edema of the face extending from within out, and rapid destruction of the soft tissues and bone with an outcome that is almost always fatal. Nowadays, cancrumoris still affects children predominantly. It occurs very rarely in the aged with decreased host resistance. Noma sequelae comprise bony and soft-tissue losses that affect both aesthetics and function. Complex defects should be treated in a staged fashion with the most complex initial stages performed in better-equipped institutions and less complex stages done on home ground. Although complete correction may be planned to restore function and aesthetics, the noma patient may eventually be satisfied with a functional but less aesthetic outcome.

Keywords: *Cancrum Oris, NOMA, Cleft lip & palate, Nasal reconstruction.*

Introduction

Cancrumoris is a disease of great antiquity described by Hippocrates and Galen. The term noma, a common synonym for the disease, is derived from the Greek verb 'to devour'. Tourdes (1948) defined cancrumoris that is still substantially correct. He described the condition as a gangrenous affection of the mouth that especially attacks children in whom the constitution

is altered by bad hygiene and serious illnesses such as exanthematous fevers. According to him, the disease is manifested by an ulcer of the oral mucous membrane, edema of the face extending from within out, and rapid destruction of the soft tissues and bone with an outcome that is almost always fatal. Nowadays, cancrumoris still affects children predominantly. It occurs very rarely in the aged with decreased host resistance.¹ Although a reconstructive surgeon may be keen to apply the principles of oncologic reconstruction to noma surgery, it must be noted that the 2 are dissimilar and a slightly different approach is necessary.² Craniofacial defects are rare, disfiguring facial anomalies, with an incidence of about to 4.9 cases per 100,000 live births. Craniofacial clefts may be caused by the failure of the maxillary processes to fuse, external pressure, amniotic bands, oligohydramnios, central disorganization of the neural crest, and hematomas.³ In 1973, Tessier classified craniofacial clefts based on clinical, radiographic, and

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surgical findings. The median cleft of the upper lip, or Tessier 0 class, has a variety of presentations, ranging from a minimal notching of the lip, vermilion, and nose to a wide cleft that divides craniofacial structures.⁴

Although great improvements have been achieved in the primary correction of the bilateral cleft lip and nasal deformity, secondary lip and nasal deformities occur in a significant number of patients. Typical secondary deformities involve broad nasal tip projection, flared alar rims, fistulae of the nostril floor, a short columella with varying degrees, and a vertically short upper lip (whistle deformity), in which severe short columella complicated with marked philtrum deficiency poses a highly demanding challenge to plastic surgeons. The columella and philtrum serve as two functional and aesthetic subunits. In deformities, a short columella often includes a deficiency of skin, nasal mucosa, and supportive alar cartilages.³ We present a case of an adult patient with noma sequelae managed by a staged approach. Although complete correction was initially planned, following the patient's requests, the last stage was abandoned and a functional but less aesthetic result accepted.

Case Report: A 55-year-old lady reported to our dept. of Oral & Maxillofacial Surgery, Institute of Dental Sciences & SUM Hospital, Bhubaneswar, Odisha, India with a large anterior maxillary facial defect involving the lips and the nose owing to cancerous about 10 years ago. The defect was large involving the upper lip in the midline, anterior hard palate, and columella of the nose (Figure 1). The surgical procedure was aimed to improve the aesthetic outcome and also increase the functional capacity of the mouth.

Defect Assessment: The defect in the present case can be compared similarly to a cleft nose and lip (Figure 2 & 3). The field of cleft surgery has seen major advances over the past 30 years. Normal function and normal to near normal appearance are a realistic goal and can be achieved.

Nasal Deformity: The abnormal anatomy of the cleft nose includes several components. The alar base, the medial and lateral crus of the alar cartilage, the nasal dome, the columella, and the nasal septum all are affected by the skeletal base, consisting of the alveolus, maxillary segments, and palate. The severity of the primary nasal deformity is intimately related to the degree of displacement, abnormality, and hypoplasia of the maxillary segments. Continued hypoplasia and

displacement of the maxillary segments, particularly the lesser segment, result in varying degrees of maxillary deficiency in the unilateral cleft lip and nose deformity. Subsequent growth and the final degree of deformity and outcome depend on the cleft dysmorphogenesis and the selection of surgical procedures and sequencing. Rehabilitation must address the three-dimensionality and growth over time to obtain the desired result.¹⁰

Lip Adhesion: A median cleft can also be called midline cleft or vertical cleft through the center of the upper lip, and is a rare anomaly, the exact developmental origin of which is not clear. Tessier 0 clefts occur during the third week of gestation due to the failure of the two medial nasal processes to close in the midline. It can occur in isolation or be a part of a syndrome such as orofacial-digital syndrome. In this case, the defect occurred due to previous aggressive infection of the anterior facial region. Various treatment options are present for mild deformities that do not involve the white roll. When developing the treatment plan, reconstruction of the cupid's bow, labial philtrum, vermilion, and buccal mucosa should be kept in mind. Some articles described using a V-Y flap, in another article they used a diamond incision to repair these anomalies. Some authors described a mucosal Z transposition technique to treat mild cases. For moderate defects involving the philtrum, Millard and Williams reported using an inverted V excision. In this case report, we have addressed both the lip component and the nasal component as a different entity to achieve maximum outcome.⁴

Surgical Technique:

Nose: The operation was performed under general anesthesia. The incision design, which involved two small triangular flaps on the nasal floor, was made along with the scarred philtral columns and extended vertically along the medial nostril walls. The prolabial flap based on the nasal tip was harvested with underlying subcutaneous tissues to protect the subdermal vascular network. By adding the alar rim incisions, an open rhinoplasty was performed. Excess fibrofatty tissue between the medial crura of the lower lateral cartilages can displace the alar domes laterally and so be removed completely. Then, the lateral and middle portion of the lower lateral cartilages was dissected from the nasal mucosa and the overlying skin. Once the desirable tip projection was obtained, the whole prolabial flap was used to cover the nasal tip and reconstruct the columella. Two accompanying small triangular flaps were inserted

carefully into the defects created by the septal mucosal contracture release (Figure 4).

Lip: Lip repair was done using the triangular flap technique initially describe by Tennison and Randall. The approach to this flap is based purely on a mathematical approach and precise measurements. The repair technique is conceptually similar to rotation advancement flap with the primary difference is that the rotation back-cut in the non-cleft side is performed more inferiorly, closer to the vermillion border. Similarly, the advancement segment on the cleft side is designed to occur inferiorly near the vermillion cutaneous border (Figure 5).



Figure 1 Defect assessment



Figure 2 Nose Defect



Figure 3 Lip Defect



Figure 4 Lip markings



Figure 5 Nasal reconstruction



Figure 6 Postoperative picture

Discussion

Noma or cancrumoris is gangrenous stomatitis affecting particularly children from developing countries. It has been known since the time of Hippocrates, Galen, and Celsus. In 1848, Tourdes used the term *cancrumoris* or *noma* for the first time and gave a clear description of the disease. In the earliest stages of the disease, a painful, red, or purple, indurated papule is present. It is most frequently seen in the posterior region of the alveolar margins. Ulceration follows rapidly and may extend into the mucosal surfaces of the cheek and the lips. Tissues appear swollen and tender to palpation. There is a profuse salivary flow accompanied by a foul-smelling and purulent discharge. Over several days, a blue-black area appears on the skin of the lips and cheeks. The gangrenous area may also involve the chin, the infraorbital region, the palate, the nose, and also the antrum. Sloughing of the tissues leaves deep, perforating wounds. The teeth are often exposed. Following the acute phase of gangrene, if the child survives, healing will proceed by contraction and fibrosis, leading to severe distortion of the mouth region.⁵

Surgery is targeted at restoring normal speech, oral competence, and acceptable cosmesis. The surgery differs from the reconstruction of defects following ablative oncologic resection in the following ways. In the adult noma patient, compound tissue losses (skin, soft tissue, bone) are commonplace. Remnant tissue is left scarred

and distorted with loss of normal anatomical reference points. Because noma occurs in early childhood, the bony defect and soft-tissue scar affect facial skeleton and dental development, producing gross asymmetry. Nutritional deficiencies are commonplace. Preoperative optimization must include nutritional assessment and correction of these deficiencies.

Complications, including poor healing and infection, are common in the immediate postoperative period. Hence, a longer observation period is necessary before discharge and return to the country of origin. Noma sequelae comprise bony and soft-tissue losses that affect both aesthetics and function. Complex defects should be treated in a staged fashion with the most complex initial stages performed in better-equipped institutions and less complex stages done on home ground. Although complete correction may be planned to restore function and aesthetics, the noma patient may eventually be satisfied with a functional but less aesthetic outcome.²

Midline defects of the face are associated with a range of developmental brain defects, trauma, or severe infectious disease leading to gangrene. Many are incompatible with life but some patients may have a normal or near-normal mentality and a full life expectancy. Reconstruction and repair utilizing existing surgical method are applicable for this latter group of patients but the midline nature of the defects may require adaptation of these techniques.⁸

The columella is an important aesthetic entity in the mid-face and following its loss or congenital absence a severe aesthetic defect results. Over the years a variety of reconstructive techniques for the columella have been described in the literature including alar margin flaps, the internal nasal vestibular flap, nasolabial flaps, and nasomalar flaps.⁸

The basic principles of treatment of the established case of Cancrumoris have remained the same. These are to release the ankylosis and only then to carry out reconstruction of the facial defect. To undertake the latter before achieving the former is disastrous for the patient, who previously was able to feed through the hole in the cheek, but with the closure of the defect is prevented from taking anything but fluids.⁷

The first step of nasal reconstruction is a thoughtful analysis of the defect. The best surgical option will provide the patient with an excellent aesthetic result and nasal function. Depending upon the extent of the defect

as well as the anatomic site, a local flap, full-thickness skin graft, composite graft, or interpolated flap will provide the optimal result. The decision will depend on the surgeon's experience and expertise, as well as the expectations and desires of the patient. Many cutaneous defects will require not only reconstruction of the defect, but also cartilage grafts to provide nasal contour and support. Most large or complex defects will require a paramedian forehead flap for reconstruction along with cartilaginous and/or bony support, as well as a lining flap.⁶

One of the most distinct problems of secondary bilateral cleft nose deformity is a short columella with a flattened nasal tip. There have been many attempts to perform columella lengthening and nasal-tip elevation. Various techniques, such as a central V-Y advancement flap from the upper lip, and bilateral forked flaps, have been commonly used to lengthen the columella. Meanwhile, different bone or cartilage grafts can be used to elevate the nasal tip and stabilize the columella. However, the most important procedures that contribute to the long-term stability of columella length and nasal tip projection are the full release of nasal mucosa contracture and sufficient soft tissue coverage at the nasal tip.

In our approach, the total height of the prolabium was used to lengthen the columella and provide adequate skin coverage for the nasal tip. Furthermore, any contracting forces induced by inadequate mucosa were fully released and the resultant defects were repaired by the two accompanying triangle flaps in the prolabial flap.³

Conclusion

Noma sequelae comprise bony and soft-tissue losses that affect both aesthetics and function. Complex defects should be treated in a staged fashion with the most complex initial stages performed in better-equipped institutions and less complex stages done on home ground. Although complete correction may be planned to restore function and aesthetics, the noma patient may eventually be satisfied with a functional but less aesthetic outcome.

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