CASE REPORT

OROFACIAL CLEFTS; A CASE REPORT OF RARE TESSIER CLEFT 0

Usman Ul Haq[1], Waheedullah Khan[2], Azhar Bangash Khan[3], Sana Tariq[4]
[1]Maxillofacial surgery department, Wah Medical College Wah Cant, [2] Orthodontics Department, Dental College HITEC-IMS, Texila, [3]Professor of Orthodontics Commandant 30 MDC Peshawar Cant, [4]Registrar Orthodontics Department of Orthodontics, Dental College HITEC-IMS, Texila.

ABSTRACT
Orofacial clefts (OFCs) represent a common but complex congenital disorder, that significantly impacts the lives of affected individuals and their families. This study probes into the complexities of OFC management, with specific focus on a rare Tessier's Cleft 0 case in a Pakistani child. The case emphasizes upon the challenges faced in low and middle-income countries (LMICs) due to limited specialized care available.
Main aim of treatment includes profile correction, maxillary and mandibular deficiency reconstruction, nasal bridge reconstruction, and achievment of functional and aesthetic dentition. Treatment alternatives cover simultaneous palatoplasty and alveoloplasty, nasal reconstruction and early maxillary protraction therapy through iliac bone graft.
This case study focuses on the importance of a multidisciplinary approach, highlighting the Tessier classification system's utility in guiding treatment decisions. Effective communication and teamwork are important, especially in LMICs where advanced care resources are limited.
Keywords: *Orofacial clefts, Tessier's Cleft 0, multidisciplinary care, low and middle-income countries.*
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INTRODUCTION

Orofacial clefts (OFCs) are one of the commonest congenital abnormalities, with a significant impact on the affected persons' survival, overall general health, development, and quality of life[1]. These patients along with their families undergo a long, seemingly never ending suffering of a tedious phase of treatment, even in cases where early specialized care is not only available but is accessible as well[2,3]. In low & middle income countries (LMIC)[4] like Pakistan, the estimate of the burden of disease, risk factors, health care provision, availability of optimum facilities and long-term prognosis with the provided care hold a key locus in decision making.

Because of the variety of presentations and rarity of the OFCs, there are complicated aesthetic and functional dilemmas that call for specialized, frequently interdisciplinary care. Executing a therapy that has been carefully planned can lessen complications and reduce the need for revision operations[5]. As we go into a preventive medicine era[1,3], it is crucial to be able to gauge the effectiveness of interventions, like surgery and rehabilitation, and correlate it with the accessibility of complex tertiary care facilities with qualified staff. This often is a difficult scenario to be found in LMICs. Similar scenario was faced by our institute, as a patient with rare OFC was referred for provision of tertiary care to our department of Orthodontics. Details of this case are reported here to highlight the complexities of disease, its treatment and management in of these patients in a peripheral dental hospital.

CAPSULE SUMMARY

A patient with the rare Tessier Cleft O is being reported. Such cases require a well-coordinated team approach besides tertiary care facilities and high clinical acumen of treating physicians. Treatment objectives are discussed.

ETIOLOGY AND DIAGNOSIS

A young Punjabi origin Pakistani boy, aged 8 years, reported to the "Maxillofacial Surgery Clinic" at Wah Medical College, Wah Cant, who was later referred to Department of Orthodontics, Dental College HITEC-IMS, Taxila. He was initially referred by a UK based NGO/ trust that provides free surgical treatment to cleft patients throughout Pakistan.

Parents of the patient presented with the primary complaint that their son's nose was deficient and had cleft inside mouth.

There was no other family member with facial or oral cleft, or any other syndromic condition. Parents had consanguineous marriage and belong to same family lineage. He had global developmental delay, poor growth, generalized weakness

Correspondence to: Sana Tariq, BDS, FCPS(Orthodontics)
Email: sana-tariq-2010@hotmail.com

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and dragged left foot while walking. Cognitively, he did not follow commands and was slow at comprehension. Speech was impaired with recurrent nasal emission of air i.e., marked velopharyngeal insufficiency. Patient had undergone palate repair by the same NGO's free cleft camp at the age of 1 year. Still patient sometimes complained of water coming through nose.

Socioeconomic status evaluation revealed that the patient belonged to a middle class income group as per Kuppuswamy scale[6].

On examination performed by pediatrician, patient's echocardiography revealed patent foramen ovale (PFO). Computed Tomography (CT) scan of the brain was performed that revealed benign enlargement of arachnoid spaces in frontal area.

His extraoral findings revealed concave profile, collapsed nose due to deficient nasal bridge but intact soft tissue structures of nose, hypoplastic maxilla and deficient malar prominences (Figure 1). Intraorally, he had deciduous set of dentition with missing deciduous central incisors (#51, #61), poor oral hygiene and multiple carious teeth. Mesial step occlusal scheme was present. There was median alveolar and palatal cleft present.

CT scan was performed to further evaluate the extent and nature of the cleft defect. CT scan revealed a cleft defect in midline. Cleft started anteroposteriorly, from alveolus in place of deciduous centrals towards the posterior limit of hard palate and vertically up towards the nose, involving base of nose that was entirely absent. All the cartilaginous structures of nose were absent whereas bony part of nasal bridge was present but not fully developed. Nasal bridge was totally collapsed that provided no support to intact soft tissue structures of nose (Figure 2).

Diagnosis of Non-syndromic isolated Orofacial cleft was made and classified as Tessier's Cleft 0.

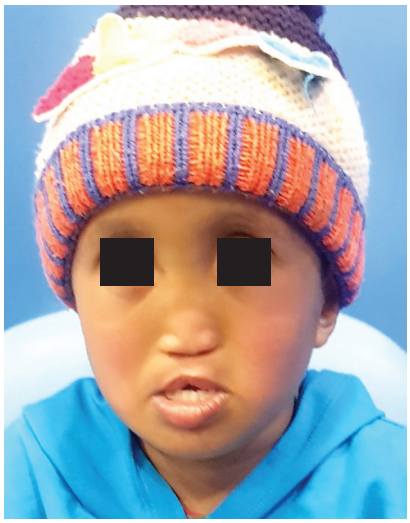


Figure 1:Extra-oral pictures.
A-frontal view

B- Profile view

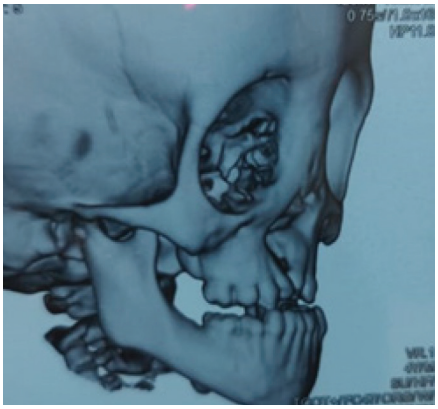
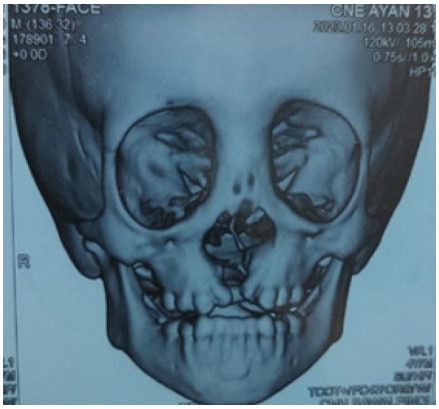
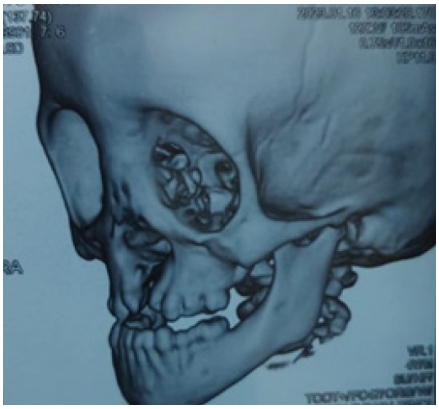


Figure 2 - CT Scan Images.
A- Right Profile view



B- Frontal View



C- Left Profile view

TREATMENT OBJECTIVES

Following treatment objectives were laid for this patient after discussion in Ortho-surgery conference of our institution.

1. Correction of profile deformity
2. Reconstruction of nasal bridge
3. Correction of maxillary and malar deficiency
4. Achievement of functional and aesthetic dentition
5. Replacement (prosthetic/natural) of missing teeth

TREATMENT ALTERNATIVES

Step wise treatment timeline was decided for this patient as follows:

Initially, simultaneous palatoplasty and alveoloplasty is planned that will include bone grafting in alveolar and palatal cleft. This procedure would be followed by early maxillary protraction therapy using facemask at around 6 years of age. Alternatively, early maxillary distraction osteogenesis with high Leefort I or Leefort II corticotomy, is also under consideration in place of facemask therapy.

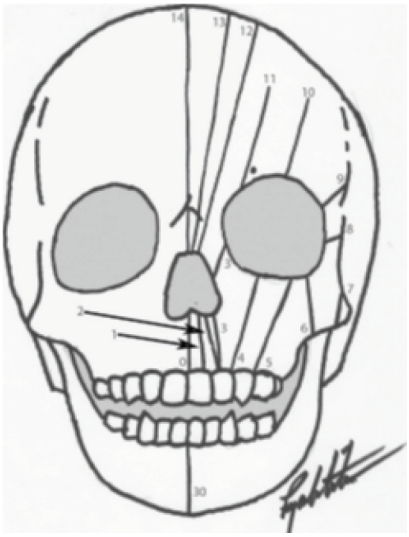


Figure 3 - Tessier Classification of OFCs

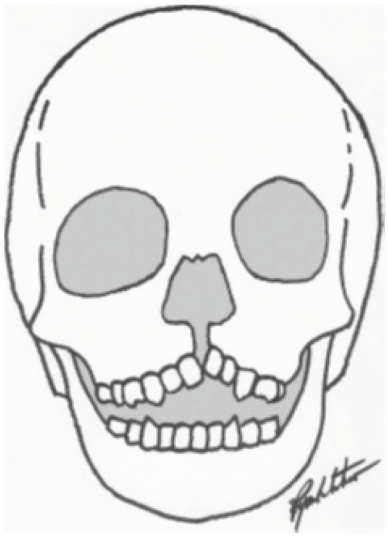


Figure 4 - Tessier 0

At a later surgery, nasal reconstruction by iliac bone graft is planned to be done. Fixed appliance treatment for correction of denture problems will be planned later depending upon how the intraoral conditions unfold.

DISCUSSION

OFCs usually present as a therapeutic challenge in terms of aesthetics as well as form and function. Treating such cases require a well-coordinated team approach besides tertiary care facilities and high clinical acumen of treating physicians.

Tessier classification[7] is widely used to classify OFCs (Figure 3) and a guidance towards a unified treatment. Our patient fell under Tessier 0 cleft (Figure 4), it is an exact midline facial cleft, may be accompanied by a Tessier 14 cleft (extension of the midline cleft to the cranium), with hypertelorism of a variable degree. Despite being one of the more frequent "atypical facial clefts," which have a frequency of 1 in a million live births, it is still uncommon[8]. Having said that, these defects are not very straight or easy to treat. A thorough and professional teamwork is required for that matter. We also planned this case after multiple discussions and re-discussions among our team and also seek advice from distant consultants. Hereafter we will be providing rationale for adopting this very treatment plan.

Simultaneous palatoplasty and alveoloplasty is considered in anticipation of development of future openbite[9] and vertical defect nearby cleft region. Putting alveolar bone graft might lead to elimination or reduction in intensity of vertical defect created adjacent to cleft.

Maxillary protraction is seemed to be necessary as profile concavity indicates presence of severe maxillary and malar hypoplasia. This is compounded by presence of mesial step occlusal scheme present intraorally, in contrast to normal i.e. flush terminal plane. All these extraoral and intraoral indicators point towards a future skeletal class III malocclusion that too be very severe in intensity. Ideal time to carry out maxillary protraction is round about 8 years. However in this case we are planning to undertake early maxillary protraction in anticipation that depressed nose and deficient malar prominences may slide along with the protracting maxilla as circummaxillary suture ossification may be at very initial stages. To our comprehension such effect may reduce the need of malar augmentation and also decrease the complexity of rhinoplasty for reconstructing the nasal projection. For similar reason, instead of using facemask we are considering placing extraoral distractors to protract maxilla. Another rationale for considering distraction osteogenesis is that patient is slow in following commands and is highly non-compliant towards treatment. Hence not a very good response is anticipated from a compliance dependent facemask appliance.

Ultimately, the reconstruction of nasal floor and reestablishment of nasal projection is under thought, as this was the chief complaint of the parents and also the reason for referral to us. Results of earlier done palatoplasty are anticipated to provide the foundation upon which nasal floor is to be reconstructed. Or if at all there still remains a need to reconstruct the nasal floor, or is it well supported by bone placed during palatoplasty. Following Ghareeb et al. strategy for nasal reconstruction we planned an iliac bone graft to create nasal height and projection[10].

Later as the patient approaches towards permanent dentition, how it unfolds? what is the intensity of anticipated vertical openbite defect? how molars and canines settle? and amount of overjet we have at that time will be the deciding factors for planning fixed appliance orthodontics.

As we have planned the case, we expect no severe skeletal anteroposterior or vertical deformity present at adolescence that may later require orthognathic surgery (OGS). Nevertheless, if things don't go as planned, a room for reconsidering the final plan or orthodontics and OGS always exists.

CONCLUSION

Contrary to the typical cleft lip, nose, and palate, the congenital atypical nasal clefts are extremely uncommon and exhibit a variety of anatomical aberrations; and in order to treat, they require intensive clinical knowledge, extensive professional experience, and an effective team effort. Good and effective communications among team members, deliberated discussions on treatment protocols and possible outcomes are essentials towards success of such cases. In LMICs where advanced care is deficient, teamwork and thorough deliberation is required to bridge the gap of lack of optimum facilities required for these complex scenarios.

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