

MISSED DIAGNOSIS OF ISOLATED BILATERAL TRANSVERSE FACIAL CLEFT: A CASE REPORT

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ABSTRACT

Introduction: Bilateral transverse facial cleft is the most common of the rare facial clefts and early presentation is a key element for successful management and prevention of possible complications like poor esthetics, speech and eating difficulties. Though several studies have documented reasons for late presentation, none has highlighted non-referral due to missed diagnosis by healthcare workers as a reason.

Case Report: A nine-year-old girl was brought by her parents to the Primary Oral Health Clinic on account of “very wide mouth” noticed at birth. The mother noticed the anomaly few hours after she gave birth to her and immediately pointed the attention of the nurses and birth attendants to it but they dismissed her concern. Subsequently, the mother took the girl to the maternity centre for routine immunization appointments, but none of the healthcare workers she encountered recognized the birth defect. The parents further reported that the girl received jests and abuses from her peers. A diagnosis of Isolated Bilateral Tessier number 7 cleft was made based on clinical examination findings. She was referred to a cleft centre where the repair was successfully carried out at no cost to the patient through the Smile-Train® sponsorship program. Subsequent follow-up visits to the primary healthcare clinic in the sixth and ninth month post-surgery revealed remarkable improvement in both patient’s and parents’ self-reported psycho-social wellbeing.

Conclusion: This case report presents a rare presentation of delayed isolated congenital bilateral macrostomia because of healthcare workers failure to diagnose.

Keywords: Congenital macrostomia, Tessier 7, Rare facial cleft, Missed diagnosis

INTRODUCTION

Congenital macrostomia also described as a Tessier number 7 cleft is a rare facial cleft¹. It usually occurs from the lack of fusion of the maxillary and mandibular processes resulting in a cleft at the commissures of the lips. While a sulcus at the commissure may be the only finding¹, it typically extends about 1 – 2 cm in length to the anterior border of the masseter², while more severe types can extend as far as the ear³ and beyond⁴. It may either occur in isolation or as part of a syndrome in which case there would be other manifestations⁵. The reported incidence ranges from 1 in 60,000 to 1 in 300,000 live births¹.

Early presentation for treatment is a key element for successful management of clefts^{6,7}. However, in developing countries, patients with clefts often present late for treatment with resulting adverse effects on the

patients’ psycho-social health and treatment outcomes^{6,7}. Studies have shown that the reasons for late presentation of these patients for care include poverty, lack of awareness and fear of treatment^{6,7}. However, none of the studies reported non-referral by healthcare workers as a reason for late presentation of patients. Hidden clefts such as isolated clefts of the palate are often missed by healthcare personnel⁸, but we did not find any report of healthcare personnel missing an external cleft such as this case of congenital macrostomia.

In this case report, we describe a case of bilateral transverse facial cleft, which was noticed by the child’s parents at birth but dismissed by healthcare personnel despite repeated attempts by the parents to get treatment for their child.

CASE PROFILE

A nine-year-old shy and withdrawn Yoruba girl, accompanied by her parents, presented at our Primary Oral Health Clinic in a rural community in Oyo State, Southwestern Nigeria on account of “very wide mouth” noticed from birth. The mother who noticed the deformity shortly after birth in a community health centre said she pointed the attention of the midwife and birth attendants to it at the time, but they simply dismissed her observation saying it will heal up or fuse together as the baby grows older. This however was not so as the deformity became wider as the child grew.

The child’s perinatal history was uneventful. The mother received regular antenatal care and delivered her uneventfully at the maternity centre. She also recorded normal developmental milestones and received a complete course of the routine immunizations. The child had not had any problems with oral functions (speech, mastication), but aesthetics had been a major

challenge. Her peers made unsavory remarks about her especially in her school. Her parents believed this had been a source of psychosocial challenge for her especially as she grew older. She showed an increasing loss of enthusiasm towards going to school and her parents had to change her school at a time. The aesthetic and psychosocial challenges were the primary reasons the parents presented at our primary oral health clinic with her.

The child is the first of two of her mother in a polygamous family of five children. The family belongs to the lower socioeconomic class; the mother who’s the first wife is a petty trader with no formal education, while the father only completed secondary school and works as a roadside mechanic.

On examination, there was an abnormal extension of the lip commissure bilaterally, more marked on the left (about 2cm) than the right (about 0.5cm). Neither extension reached as far as the anterior border of the



Fig. 1: The Lateral (left) and facial views of the girl at presentation.



Fig. 2: Immediate post-op facial pictures of the girl.



Fig. 3: The lateral and frontal views of the girl at nine (9) months post-op.

Masseter muscle. However, an abnormally wide mouth opening was present (Figure 1- pre-op photo). No evidence of any associated deformity nor associated syndromes was noticed, hence a diagnosis of Isolated Bilateral Tessier Number 7 cleft (Grade I) was made based on the physical examination findings ⁹.

The patient and parents were then counseled about the defect. The patient was subsequently referred to a specialist hospital in the capital city of Ibadan, southwest Nigeria for surgical repair of the deformity. The option of repair was readily and happily accepted not only because it would alleviate the attending challenges, but also because it would be at no cost to the parents since it was sponsored by a non-governmental organization (Smile Train). The surgery was successfully carried out (Figure 2- immediate post-op photo). She was reviewed at 1 week, 1 month, 3 months, 6 months, and 9 months post-op, and the reviews were uneventful. At the latest review (Figure 3 – 9 months post-op photo), she was cheerful and reported that she was happy at school. The parents were also satisfied with the outcome of the surgery and expressed their satisfaction with her appearance and newfound enthusiasm for school.

DISCUSSION

This report described a girl in a rural area in Nigeria who was denied access to care for bilateral congenital macrostomia for nine years because the healthcare personnel that she came in contact with up to that point did not recognize congenital macrostomia. This highlights a possible gap in the knowledge of healthcare personnel in rural areas about congenital

macrostomia and possibly other rarer facial clefts especially when they are not the severe form. It was disheartening to find that only 5 (less than 20%) of the 26 cases of isolated congenital macrostomia reported in the literature presented for care in the first year of life ^{1,5,10-13}.

The reasons proffered for this late presentation include the fear of stigmatization¹⁰, lack of awareness, poverty, cultural beliefs, lack of access to appropriate health facilities ^{6,7,14}, and poor referral systems ¹⁴. The case of this nine-year-old girl is therefore significant because it highlights a never before reported aspect of “ignorance”. The ignorance of healthcare workers. Previous reports show that healthcare workers often miss hidden defects such as clefts of the soft palate ⁸, but the cleft we report was on the face.

Unfortunately, due to the delay, she was already experiencing some of the reported social complications like “social anxiety”¹¹. This was evidenced from the report by her mother that she was becoming increasingly “shy and withdrawn” especially when she is with her peers. All the other associated negative consequences that she was reported to have passed through, like the loss of enthusiasm to go to school, leading to change of schools, poor academic performance, and low self-esteem show that even when the facial cleft did not limit function such as eating and talking, it has the potential of causing devastating social problems if not repaired early enough.

Clefts are managed surgically by specialists in urban areas at prohibitive costs. However, organizations like

Smile Train have provided financial access to cleft services for over ten years¹¹, and it is therefore tragic that this girl was denied access to treatment for so long.

CONCLUSION

This case report presents a rare presentation of delayed isolated congenital bilateral macrostomia because of healthcare workers failure to diagnose. Therefore, further studies are needed to objectively assess the knowledge of healthcare workers on congenital birth defects. Furthermore, continuous medical education for all cadres of healthcare workers on the identification, diagnosis and prompt referral of patients with congenital birth defects should be instituted.

CONFLICT OF INTEREST

The authors declare no conflict of interest exists.

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