

CASE REPORT:
**A TRUE MEDIAN FACIAL CLEFT (CRANO-FACIAL DYSRAPHIA, A TESSIER TYPE O) IN
BINGHAM UNIVERSITY TEACHING HOSPITAL, JOS**

* **GARGADI S I, ** JIMOH A O**

* PLASTIC & RECONSTRUCTIVE SURGEON BINGHAM UNIVERSITY TEACHING HOSPITAL
JOS PLATEAU NIGERIA (BHUTH)

** PAEDIATRICIAN BINGHAM UNIVERSITY TEACHING HOSPITAL JOS PLATEAU NIGERIA

*Corresponding Author: Gargadi SI,
Plastic & Reconstructive Surgery Department
Bingham University Teaching Hospital Jos
Email: drgargadi@yahoo.com
Phone: +234 8033155011*

ABSTRACT

A female Newborn given birth to by a 25years old P³⁺⁰ 3alive via spontaneous vaginal delivery after 38 weeks of supervised normal pregnancy in Bingham University Teaching Hospital (BHUTH) Jos with a true Median Facial Cleft (Cranio-facial dysraphia, a Tessier type O). She was in stable state with occipitofrontal dimension of 38cm, posterior sutural dimension of 6x10cm and epicanthic distance of 15mm. She had an upper lip midline defect, absent columella and anterior nasal Septum.

Cranio-facial dysraphia is rare with incidence of 1.4-4.9 per 100,000 live births. A True Median Cleft of the face, occurs due to genetic and environmental causes because of delay in the closure of the anterior neuropore. Two of the paternal cousins of the patient had cleft lip which supports a genetic predisposition.

Patient had a multidisciplinary care by the obstetrician, Neonatologist, anesthesiologist and the plastic surgery team who scheduled a soft tissue repair of the upper lip defect, columella and anterior nasal septal defect. However, the patient died of febrile illness before surgery

KEYWORDS: *True Median Facial Cleft, Crano-Facial Dysraphia, A Tessier Type O*

INTRODUCTION

In Pre-Hispanic American history 200AD-900AD 3 vessels were found in Monche burials pottery in Peru with median Cranio-facial dysraphia associated with midline cleft of the lower lip and mandible.¹ Median cranio-facial dysraphia occurs due to failure or delay in the closure of the anterior neuropore.² There is failure of neuroectodermal migration and penetration, the epithelium breaks down to form facial cleft. Dursy in 1869 and His in 1892 purported the failure of fusion of facial processes by mesenchymal penetration results in craniofacial anomalies.³

The median craniofacial dysplasia has tissue agenesis and holoprosencephaly at one end (the hypoplasias), frontonasal hyperplasia and excessive tissue (the hyperplasias) at the other end, and abnormal splitting or clefting and normal tissue volume (dysraphia) occupying the middle portion of the spectrum.⁴

Paul Tessier classify craniofacial clefts into oral-nasal clefts: 0,1,2,4, oral ocular clefts: 4,5,6, Lateral facial clefts: 7,8,9, cranial clefts:

10,11,12,13,14.^{5,6} American Association of cleft palate Rehabilitation (AACPR) Classify cranio-facial cleft into: (1) Mandibular process cleft, (2) Naso-ocular clefts, (3) Oro-ocular clefts and (4) Oro-aural clefts.³ Estimated incidence of Cranio-facial cleft incidence is 1.4-4.9 per 100,000 live births which is rare compared to cleft lip and palate.³ Rare type occurs sporadically, role of heredity in causation occurs in cranio-facial clefts seems to occur in Treacher-Collins syndrome and in some familial cases of Goldenhar syndrome. A dominant gene defect (TCOF-1) causes Treacher-Collins syndrome. Environmental causes include infections such as TOUCHES syndrome, maternal metabolic imbalances, drugs and teratogenic chemicals.³ It has also been reported to be produced by disturbances of embryogenesis from repeated hypoxia.⁷ The index patient present with a True Median cleft with no hypoplasia or hyperplasia (A Type O Tessier's classification).

(A)



(B)



Fig. 1: (A) show an anterior view, (B) Show a side view of a female new born with a true midline facial cleft (Tessier type 0)

CASE REPORT

A female New born given birth to by a 25years old P³⁺⁰ 3alive housewife via spontaneous vaginal delivery after 38weeks of pregnancy that was supervised at the Jos North primary health Centre. Pregnancy was uneventful, and baby cried immediately after birth. Mother had minor perineal laceration which was repaired in our facility where

she delivered. Two of the father's cousins had cleft lip which was repaired at childhood.

The female Newborn was noticed to be active, Pink, a cyanosed, anicteric, well hydrated with Heart beat of 136 beats/min, Heart sounds I and II only, respiratory rate of 36 circles per minute, Temperature of 36.1°C and SPO₂ was 96% in room

air. She weighed 3.2kg, occipitofrontal head dimension was 38cm, posterior sutural dimension was 6cm x 10cm and epicanthic distance 15mm. She had a midline defect in the upper lip extending to the gingiva, absent cupid bow, absent philtra ridge and groove, absent columella and absent anterior nasal septum. No other congenital anomalies were present.

Her pack cell volume was 46%, total bilirubin was 7.3mg/dl (normal is < 12mg/dl). She was scheduled to have Millard's repair with "Forked" flap and nasolabial Island flap to repair the columella and the anterior nasal septum however the patient died before the surgery date from a febrile illness.

DISCUSSION

Cleft lip and palate is the most common cranio-facial anomalies with incidence of 1:2000 in Africa, 1:1,000 in Caucasians and 1:500 in Asia.^{2,8} Cranio-Facial dysraphia has an incidence of 1.4-4.9 per 100,000 live births which makes it rare and reportable when it occurs in our health facility. Congenital anomaly of cranio-facial has variable expressivity or multifactorial suggest a threshold over which the cleft phenotype is expressed. Genetic or environmental factors may carry one over the threshold.⁷ The risk of having cleft lip and palate when one sibling is affected is 4%, for two siblings affected the risk is 9%, and when one sibling and one parent are affected the risk is 17%.² In the index patient two of the paternal cousins had cleft lip which suggest a genetic predisposition.

The patient was scheduled to have soft tissue repair of the median cleft defect using Millard's repair with Lateral flaps elevated as "forked" flap plus nasolabial flap to reconstruct the upper lip midline defect, columella, and anterior nasal septum however the patient died of febrile illness before surgery.

CONCLUSION

The occurrence of a true median facial cleft in BHUTH Jos Plateau Nigeria, a rare cranio-facial anomaly is worthy of observation.

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