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Case Report

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Cranio-Facial Duplication (Diprosopos) with Myelomeningocele

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ABSTRACT
Diprosopos is an extremely rare form of congenital anomaly that leads to complete or incomplete duplication of cranio-facial structures. Majority of diprosopos are females and delivered as stillborn with few surviving not longer than neonatal period. This report aims to describe a rare occurrence of diprosopos with myelomeningocele in an unbooked mother and recommend antenatal care and regular obstetric ultrasounds for early diagnosis and management decisions on this uncommon congenital anomaly. We report a case of a female neonate with complete duplication of the face (diprosopos) delivered via caesarean section by a 30-year-old unbooked multipara. Child's clinical condition suddenly deteriorated and died on second day of life.

Introduction

The condition Diprosopos is derived from Greek words: di- means two, while prosopon referring to face, as such diprosopos describes a clinical condition characterised by double face in a monocephalic and single trunk individual [1,2]. Diprosopos also referred to as craniofacial duplication occurs when all or parts of structures of the face were duplicated [2,3]. It is a rare form of anomaly accounting for 10% of all conjoined twin [2-4]. Ambroise Pare´ was credited with the first report of diprosopos in 1864 and since then, reports continue to be published on this rare congenital anomaly [5].

Diprotodons is an extremely rare pathology with a reported incidence of 1 case in 180,000-15,000,000 births [3,5,6]. Recent systemic review of this condition revealed only 40 reported cases globally [6]. The exact aetiology and pathogenetic mechanism explaining the development of craniofacial duplication (diprosopos) is not known but likely role of sonic hedgehog (Shh) receptors on embryonic development of craniofacial tissues and differentiation has been elucidated [7]. The risk factors for craniofacial duplication identified include advanced maternal age, polyhydramnios and consanguinity and other predisposing factors associated with conjoined twins include underweight mother and abnormal calcium metabolism [8].

Craniofacial duplication has been classified by Gorlin and colleagues in 1990, into single mouth with duplication of the maxillary arch, supernumerary mouth laterally placed with rudimentary segments, single mouth with replication of the mandibular segments, and true facial duplication, called diprosopos.

Years before Gorlin's classification, Barr in 1982, categorised craniofacial duplication into:

- Duplication of the eyes and nose with or without maxillary duplication by itself or with mandible duplication
- Duplication of the nose with or without maxillary duplication
- Duplication of the maxilla with or without mandible or pituitary duplication [1-10].

The management of craniofacial duplication depends on the severity of duplication, cases with fewer duplicate structures are easy to evaluate and manage and have better outcome compared to complete facial structural duplication in diprosopos. Craniofacial imaging including ultrasound, computerised tomography scan and magnetic resonance imaging help in assessing the extent of craniofacial duplication, defects and associated anomalies. Pre-operative evaluation of craniofacial duplication assists in surgical planning and prediction of management outcome.

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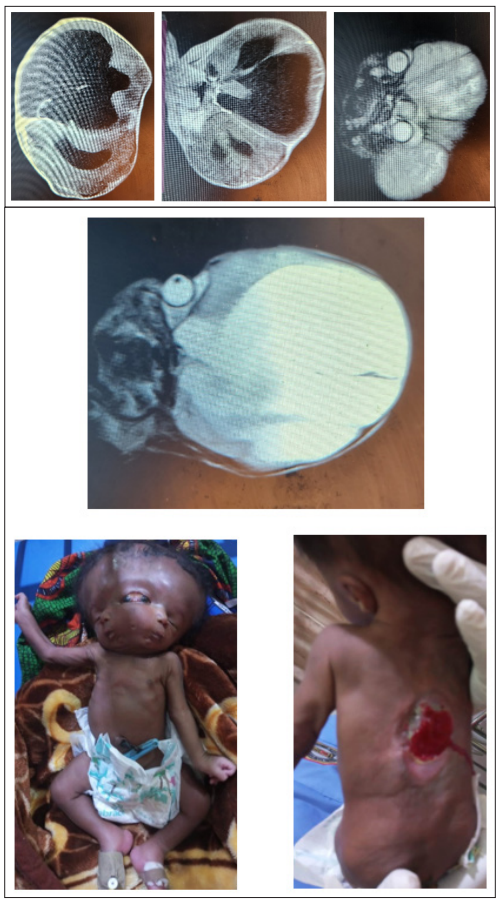
Most cases of diprosopos do not survive neonatal period with significant cases being stillbirth [4].

This report aims to describe the clinical and radiological findings of a neonate with an uncommon complex craniofacial duplication and contribute to the pool of literature on this rare congenital anomaly.

Case Presentation

A day-old female neonate was referred to our neurosurgical centre from a rural hospital with an enlarge head, duplications of all facial structures including forehead, both eyes, nostril, mouth, maxilla and mandible (figure 1). The neonate was delivered a caesarean section following prolong obstructed labour. Mother is a 30-year multipara with no history of antenatal care or similar condition in the family. There was history of maternal febrile illness treated with antimalarials and paracetamol. When examine, we found a lethargic neonate with one enlarged head with 2 faces fused at the midline and duplications of facial structures: with a pair of forehead, eye, nostril and mouth for each face. There were 2 intact ears. Occipito-frontal circumference was 48 centimetres, both fontanelles were patent, single trunk and normal looking both upper and lower limbs. Chest was clear clinically and abdominal examination revealed normal findings. Child had normal female external genitalia. Cranio-facial magnetic resonance revealed marked dilatation of two lateral ventricles, two separate hemispheres containing separate ventricles, agenesis of corpus callosum, encephalomalacia and two joined eyes with separate eye balls. The child was resuscitated with fluids and had vitamin K administered, clinical condition deteriorated within 24hrs with worsening respiratory distress and died two days after birth.



Discussion

Craniofacial duplication anomalies are extremely uncommon globally. Few cases were reported in Nigeria and other Sub-Saharan African countries. Most of the reports in Nigeria were incomplete facial structure duplication with the index being really rare especially when its complex associated with other central nervous system anomalies. The prevalence rate of diprosopos is 2 per 1,000,000 births, this makes craniofacial duplication rare as having less than five cases per 10,000 [2,11].

The present case was recorded in a female neonate, as reported in several published data, female preponderance was observed, with a male-to-female ratio of 1:1.4 [2]. Among published articles few cases were seen in males compared to female gender. Similar to parasitic craniopagus, which craniofacial duplication was conceived to be related to, also, have female preference [8].

The index case was a female neonate with duplication of both eyes, foreheads, nostrils and mouth. Joined at the midline with single head. There was an ulcerated myelomeningocele extending from lower thoracic to lower lumbar spine area. Cranial magnetic resonance imaging showed single cranium with two separate dilated ventricles and poorly developed corpus callosum. There was no history of consanguinity or similar occurrence in the family. Complete craniofacial duplication was found to be associated with central nervous system, respiratory, cardiovascular and gastrointestinal anomalies [1,12]. In the current presentation, only central nervous system anomalies such as hydrocephalus, myelomeningocele were observed. No other anomalies found. Possibility of occurrence of other anomalies involving many body systems mentioned earlier requires further evaluation such as echocardiography for heart anomalies, abdomino-pelvic ultrasound or magnetic resonance imaging for gastrointestinal anomalies and chest radiograph or computed tomography scan. Where multiple anomalies involving different system, multi-disciplinary approach to management is required to improve care and survival of patients with this kind of congenital anomaly. Treatment of incomplete duplication of facial structures was easier and associated with fewer complications compared to cases bedevilled by complete craniofacial duplication. Conditions with incomplete craniofacial duplication is treated by excision of the duplicated facial structure when found safe and feasible. Complete craniofacial duplication is difficult to treat and usually associated with poor outcome and many do not survive longer than neonatal period.

Conclusion

This report describes an uncommon occurrence of complete craniofacial duplication with thoraco-lumbar myelomeningocele who succumbed few days after birth and added to the pool of literature on diprosopos anomaly.

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