DOI: 10.5958/j.2319-5886.3.2.095

International Journal of Medical Research & Health Sciences

www.ijmrhs.com Volume 3 Issue 2 (April - Jun) Coden: IJMRHS Copyright @2014 ISSN: 2319-5886

Received: 30th Dec 2013 Revised: 25th Jan 2014 Accepted: 30th Jan 2014

Case report

FETAL DIPROSOPUS (DOUBLE FACE): A CASE REPORT

*Onankpa BO[1], Ukwu E[2], Singh S[2], Adoke AU[2], Tahir A[1]

[1]Department of Paediatrics, Usmanu Danfodiyo University Teaching Hospital, PMB 2370, Sokoto, Sokoto State, Nigeria.
[2]Department of Obstetrics and Gynaecology, Usmanu Danfodiyo University Teaching Hospital, PMB 2370, Sokoto, Sokoto State, Nigeria.

*Corresponding author email: benonankpa@yahoo.com

ABSTRACT

Diprosopus is an extremely rare form of congenital anomaly that results in partial or total duplication of the face. Most cases of diprosopus are delivered as stillborn or die few moments after delivery. The aim of this report is to alert clinicians that the antenatal finding of polyhydramnious may be strongly associated with fetal diprosopus, this routine high resolution anomaly scans should be recommended to help detect such anomaly early in pregnancy. We report a case of a female neonate with partial duplication of the face (diprosopus) delivered by a 39 year old booked multipara. Baby's condition deteriorated within 24hrs with worsening respiratory distress and died on the 2nd day of life.

Key words: Facial diprosopus

INTRODUCTION

Diprosopus, is a Greek word for two-faced.[1] This congenital anomaly is often referred to as craniofacial duplication in which there is partial or total duplication of the face. However, the fetus has a single trunk with normal limbs. In a typical presentation the fetus has a duplicated nose with eyes spaced far apart, but, in extreme cases, the baby has the entire face duplicated (i.e. diprosopus). It is an extremely rare condition with a reported incidence of 1 case in 180,000-15 million births.[2] Cases of fetal diprosopus have been reported in Italy[3], Germany[4], Spain[5], Saudi Arabia[6], Turkey[7] and India.[8] To our knowledge this is the second case reported in Nigeria, after the case reported by Ibrahim et al[9]

Several attempts have been made by several researchers to explain the mechanisms that leads to craniofacial duplication. In the presence of two completely formed, but identical faces, the babies are often referred to as a rare variant of conjoined twins. This mechanism resulting in two faces is considered to have occurred as a result of cranial bifurcation during neurulation of the notochord. [10] Two vertebral axes develop alongside the neural plates as a result of the bifurcation including neural crest derivatives. Facts from the literature also revealed that 0.4 percent of diprosopus is seen in conjoined twins.[10] Another possibility is that there could be an increased production of sonic hedgehog (SHH), a protein which is essential for craniofacial patterning during fetal development; this has already been demonstrated in chicks by researchers in an experimental studies.[11] These studies showed that the chicks were born with anomalies including double beaks with their eyes spaced far apart. In contrast, the researchers also

found that too little SHH led to abnormal midline facial features resulting in cyclopia.[11]

Prenatal diagnosis is possible for diprosopus using in utero technics including ultrasound and computer tomography (CT) scanning. The presence of polyhdromnios is considered as a strong indication of craniofacial duplication. Presently, there is no treatment for diprosopus however, termination of pregnancy is sometimes considered an option, especially if diagnosed is made early in pregnancy.

Children with this defect are normally stillborn, but a young girl, Lali Singh, born in 2008 survived for 2 full months before dying of a heart attack. [12] Partial facial duplication, as in our case, is associated with fewer anomalies, and the prognosis is better with symmetry and an excess of tissue, rather than a deficiency, favoring a positive result. [13] Most of the cases reported were females (4 males/11 females). [5] To our knowledge, there have been less than 150 reports of diprosopus in the world medical literature. [9]

CASE REPORT

Baby AM, a female was delivered to a 39- year old booked multipara. The mother had regular antenatal visits. The mother was admitted on account of polyhydramnious at 36 weeks gestation. She had abruptio placenta while on admission and, she was delivered of the baby by emergency Caesarean section. Baby required minimal resuscitation, had Apgar scores of 7 & 9 at 1st and 5th minutes respectively. The birth weight was 2800grams, length was 47centimeters and the occipito-frontal circumference was 35centimeters. Baby was uniformly pink, in mild respiratory distress with SPO2 of 98%. The major findings were on the head and neck.

Baby had one cranium with 2 faces fused at the midline, with a pair of mouth for each face. The left face had an intact nose with the right face having a trunk like organ with a single opening for a nose. There were 2 intact ears. The left face had a well formed left eye; the other incomplete eye appeared to fuse in a transverse groove across the faces. Each head had an anterior fontanelle. Baby had normal female external genitalia. Baby's condition deteriorated within 24hrs with worsening respiratory distress and died on the 2nd day of life. The parents declined autopsy. However, we got an ethical clearance from UDUTH ethical committee and an informed consent from the parents to report the case.

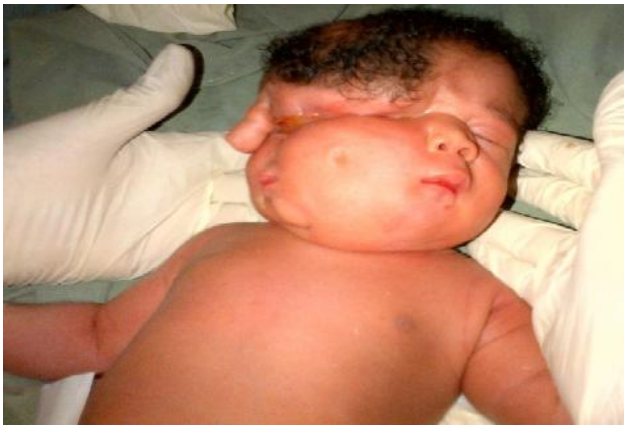

Fig 1: Female neonate with diprosopus

DISCUSSION

Craniofacial duplication remains a rare entity with only 27 cases reported since 1900.[14] Polyhydramnios, a condition that is considered as an indication for craniofacial duplication was detected in this patient. The risk factor for congenital anomaly includes familial tendencies, advance maternal age, previous history and polyhydramnious**.** The patient was of advance age and developed polyhydramnious during the third trimester. However, repeated ultrasound scan done could not reveal other findings apart from polyhydramnious. Perhaps a high resolution 4D scan would have detected these congenital anomaly and also highlight others if any. Other investigations, including CT-scan, MRI may have helped in the diagnosis, but the cost of this investigation is beyond the reach of most patients in our environment. Congenital anomalies are associated with myths and beliefs, particularly in Africa and sub-Saharan Africa. Thus, such babies are abandoned and poorly cared for by their parents. Probably because diprosopus is rare, few management options including corrective surgeries have been documented.[15] The prognosis and fetal outcome has remained unfortunately poor in all cases reported.

CONCLUSION

Diagnosis may be quite elusive during pregnancy with many cases undetected before delivery. High index of suspicion, full evaluation of cases of polyhydromnious and high resolution scan may help in the early diagnosis and prompt intervention.

Conflict of interest: None

REFERENCES

1. Al Muti Zaitoun A. Chang J, booker M. Doprosopus (Partially duplicated shead) associated with anencephaly. A case report. Pathology Research and Practice. 1999;195:45-50
2. D'Armiento, Massimo, Jessica Falleti, Maria Marilotti, Pasquale Mertinelli. Diprosopus Conjoined Twins: Radiologic, Autopic, and Histological Study of a Case. Fetal and Pediatric Pathology. 2010;29:431–38
3. Pavone L, Camera G, Grasso S, Gambini C, Barberis M, Garaffo S etal., Diprosopus with associated malformations: report of two cases. Am J Med Genet. 1987;1:85-88
4. Stefan Hähnel, Peter Schramm, Stefan Hassfeld, Hans H Steiner, Angelika Seitz. Craniofacial duplication (diprosopus): CT, MR imaging, and MR angiography findings case report. Radiology. 2003;1:210-3.
5. María Luisa Martínez-Frías, Eva Bermejo, Jacobo Mendioroz, Elvira Rodríguez-Pinilla, Manuel Blanco, Javier Egüés, Valentín Félix. Epidemiological and clinical analysis of a consecutive series of conjoined twins in Spain. Journal of Pediatric Surgery. 2009; 44(4):811-20
6. Amr SS, Hammouri MF. Craniofacial duplication (diprosopus): report of a case with a review of the literature. Eur J Obstet Gynecol Reprod Biol. 1995;1:77-80
7. Kutsi Koseoglu, Cantay Gok, Yelda Dayanir. CT and MR imaging findings of a rare craniofacial malformation: diprosopus. Am J Roentgenol. 2003;3:863-64
8. Suhil A Choh, Bakshi Jehangir, Naseer A Choh, Omar Kirmani, Roomi Yousuf. Imaging findings in diprosopus tetraophthalmos: a case report. Pediatr Int. 2010;1:54-6
9. Ibrahim A, Mshelbwala PM, Ajike SO, Asuku ME, Ameh EA. Diprosopus (double mouth) in a Nigerian child: Case report and literature review. Nigerian Journal of Plastic Surgery 2012;8(2)
10. Dhaliwal Harjit, Paul Adinkra, Diane Ennis, and Pauline Green. Monocephalus Diprosopus (Complete Craniofacial Duplication) Associated with Hydrancephaly and Other Congenital Anomalies. Priory Lodge Education Limited, 2007. http://priory.com/medicine/Birth_Abnormality.htm (Accessed November, 16, 2010).
11. Hannel, Stefan, Peter Schramm, Stefan Hassfeld, Hans Steiner, and Angelika Seitz. Craniofacial Duplication (Diprosopus): CT, MR Imaging, and MR Angiography Findings. Radiology.2003;226: 210–13
12. Jamie Frater Top 10 Bizarre Medical Anomalies. LISTVERSE. January 8, 2009.
13. June Wu, David A Staffenberg, John B Mulliken, Alan L Shanske. Diprosopus: a unique case and review of the literature. Teratology. 2002;6:282-87
14. Turpin IM, Furnas DW, Amlie RN. Craniofacial duplication (diprosopus). Plast Reconstr Surg. 1981;67: 139e-42
15. Okazaki, Joel, James Wilson, Stephen Holmes, Linda Vandermark. Diprosopus: Diagnosis in Utero. American Journal of Roentgenology. 1987;149:147–48.