Case Report

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Facial Duplication (Rare Diprosopia) about A Case

DJELTI SIHEM*, BENMOUMEN MOHAMMED YASSINE

Ehs Nouar Fadela ex st Anne Oran Algeria

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Abstract

A new male born referred to our department with a total facial duplication whose maternal operative indication was for maternal medical reasons and on the basis of obstetric ultrasound which spoke of a scimois twin. Our goal of this article is to share a very rare congenital anomaly.

Keywords: Diprosopia, Craniofacial duplication, Protein anomaly, Very rare congenital malformation

Introduction

Diprosopia or craniofacial duplication is an extremely rare congenital disease (1). From the ancient Greek "di-" and "prosopon" (literally "two faces"), we speak of diprosopia when a child is born with a split face-in medical language, this is a "partial facial duplication ".

This is a rare form of conjoined twins that comes in different forms, where parts of the face or the whole face are duplicated on the head; it can be symmetrical or asymmetrical and affect the nose, maxilla, mandible, palate, tongue and mouth. It generally occurs at the same time as other congenital diseases such as anencephaly, neural tube formation, congenital heart disease (2,3). This malformation should not be confused with Siamese twins, as it is not due to the fusion or incomplete separation of two embryos, but rather the result of an abnormality in a protein that marks the normal craniofacial pattern.

Observation

A new male child from a 28-year-old mother, G2P1, with no living children and no particular medical history who presented to our department for the management of a 24-week pregnancy with amenorrhea on scarred uterus with Siamese type fetal malformation on ultrasound which was done by an expert who spoke of a two-headed parapagus which is also an extremely rare malformation. A medical termination was indicated for her due to fear of uterine rupture due to scarring of the uterus.

Previous pregnancy even at term. The child died periportunally due to undiagnosed cardiac malformation during the pregnancy. current pregnancy; her progress in the first trimester was unremarkable, an ultrasound was done by a general practitioner who spoke of a progressive monofetal intrauterine pregnancy and the rest was unremarkable, in the second trimester the patient went to a gynecologist for a possible morphogram which did a second trimester obstetric ultrasound where he discovered that there was a cervical malformation hence his referral to an expert in in morphogram which spoke of a parapagus.

The patient presented to us with anxiety, a collective decision between obstetrician gynecologist, pediatrician and intensivists for a medical termination of the pregnancy due to possible uterine rupture on scar (scar not assessed before pregnancy), and also depression.

The cesarean section was planned, for fetal extraction which was difficult, a newborn male with total facial duplication (diprosopia) (figure 1 and 2).

The new born died within minutes of giving birth. Operative sequels were unremarkable.

Discussion

This type of craniofacial duplication is very rare with approximately 35 cases described in the medical literature(3), a defect in embryonic development which can lead to a duplication of certain structures of the face or of the entire face, It is associated with other malformations(4). Our observation illustrates a rare case of living diprosope, after that of Dr. a case was described in 1972 in New Mexico (United States) with two mouths and duplication of the mandible and maxilla (5). Authors have described similar cases but these reported cases died in their first months of life (1,2).

***Corresponding author:** Djelti Sihem, Obstetrics and gynecology service: Ehs Nouar Fadela ex st Anne Oran Algeria. Faculty of Medicine Ahmed Benbella 1 of Oran Algeria; E-mail: djeltisihem@gmail.com

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Figure 1: Diprodopia newborn of 24 weeks with Amenorrhea.

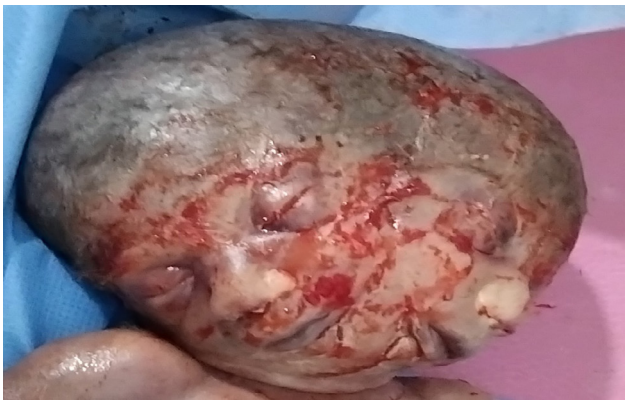

Figure 2: Facial duplication.

The incidence of this malformation is 1 in 2800 to 200,000 births (6). A female predominance has been noted in the literature (6). It is the rarest and most serious form of conjoined twins according to Mach in 1993; in the majority of cases these were stillbirths (7,8). The embryological explanation is still poorly understood.

Is the result of 'an abnormality of a protein that marks the normal craniofacial pattern, abnormal activity of the Sonic hedgehog (SHH) protein, which plays a major role in craniofacial arrangement during embryonic development. This protein is notably responsible for the width of facial features. Even if this disease is often compared to the case of Siamese twins, it is not due to the fusion or incomplete separation of two embryos. The SHH protein is notably responsible for the width of facial features. In extreme cases, these results in broadening of features and duplication of facial Conjoined twins can affect any part of the fetus's body, including the face. The case occurs as a result of a partial splitting of a developing embryo at later stages of development. The later the division occurs, the more likely it is that conjoined twins will occur (9).

The greater the enlargement, the more numerous the duplicated structures; these are often symmetrical. In the laboratory, tablets containing the SHH protein was introduced into chicken embryos: the hatched chicks had duplicate beaks. Too little of the SHH protein leads to opposite excesses such as cyclopia where facial features are not sufficiently developed.

Effective brain development also depends on the SHH protein signaling pathway. During embryonic development, the protein affects embryonic cells to specific areas which then form specialized nervous tissue: the size and shape of brain structures are therefore dependent on (9).

Most infants with diprosopia are stillborn. Few cases report humans surviving more than a few minutes or hours. However, the medical literature reports a few cases of people suffering from diprosopia who lived into adulthood, and sometimes even into old age (10).

Six year old girl with two noses and the outline of a third eye. Thus, in 1881, a French anatomist reported the case of a diprosopic woman who lived in Saint-Maigner, in the Central massif. This woman had two noses, the outline of a third eye between them, a bifid uvula and six upper incisors. Although her brain showed partial duplication, she had normal intelligence and did not die until age fifty-two (9).

Another case of a diprosopic man who nevertheless lived a long time has been described in the literature, an American circus performer who also had two noses, the outline of a third eye and a large cleft lip and palate between his two nasal appendages. A case reported in the literature, he had normal intelligence and did not hesitate to speak with the public who gazed at him. He lived for sixty-three years, from 1913 to 1976(10).

More recently, in 2002 and 2003, the medical literature reported two cases of male infants with partial diprosopia who were able to survive. However, one of them remained severely mentally handicapped, his brain having been damaged by significant cranial deformations. The last case that was described was in 2020, American ENT surgeons and pediatricians described a (very rare) case of diprosopia in the scientific journal BMJ Case Reports. It concerns a little girl: during the prenatal ultrasound of the third trimester of pregnancy, doctors first detected the presence of a "mass" in the right lower jaw of the fetus.

Our patient unfortunately had a first trimester ultrasound done by a general practitioner who was unable to identify the malformation; the duplication was across all facial structures (11). The treatment of this type of malformation remains surgical, but the prognosis is poor, especially in case of duplication complete; it is based on the elimination of the duplicated component to have the most normal appearance possible. Success of these interventions is variable (especially appearance of cystic formations reported by several authors) (12).

Conclusion

Diprosope remains a rare entity with around 35 cases described in literature. It exists in different forms: symetric (which remains a rare form) or asymmetrical, and can interest all parts of the face: the nose, the eyes, the maxilla, mouth or mandible.

References

1. Suryawanshi P, Deshpande M, Verma N, Mahendrakar V, Mahendrakar S (2013) Craniofacial duplication: a case report. J Clin Diagn Res 7: 2025-2026.
2. Onankpa BO, Ukwu E, Singh S, Adoke AU, Tahir A (2014) Fetal diprosopus (double face): a case report. Int J Med Res Health Sci 3: 461-463.
3. Borzabadi-Farahani A, Yen SL-K, Yamashita DD, Sanchez-Lara PA (2012) Bilateral maxillary duplication: case report and literature review. Oral Surg Oral Med Oral Pathol Oral Radiol 5: 29-32.
4. Costa MA, Borzabadi-Farahani A, Lara-Sanchez PA, Schweitzer D, Jacobson L, Clarke N, et al. (2013) Partial craniofacial duplication: a review of the literature and case report. A review of the literature and case report. J Craniomaxillofac Surg 42: 290-296.
5. Tripathy R, Mishra S, Sabhlok SS (2013) Accessory maxilla with a lateral cleft lip: a rare case report. J Oral Maxillofac Surg Med Pathol 25: 238-240.
6. Pengiran Suhaili DN, Somasundaram S, Lau SH, Ajura AJ, Roslan AR, Ramli R (2011) Duplication of lower lip and mandible: a rare diprosopus. Int J Pediatr Otorhinolaryngol 75: 131-133.
7. Hahnel S, Schramm P, Hassfeld S, Steiner HH, Seitz A (2003) Craniofacial duplication (diprosopus): CT, MR imaging, and MR angiography findings case report. Radiology 226: 210-213.
8. Stiehm WD. Facial duplication (1972) Am J Roentgenol 116: 598-601.
9. Hegazy A (2014) Clinical embryology for medical students and postgraduate doctors. Lap Lambert Academic Publishing.
10. Chowdhury SR, Roy A (1991) Duplication of the upper lip and maxilla. Br J Plast Surg 11: 468-469.
11. Avery JK, Hayward JR (1969) Case report: duplication of oral structures with cleft palate. Cleft Palate J 6: 506-515.
12. Hähnel S, Schramm P, Hassfeld S, Steiner HH, Seitz A (2003) Craniofacial duplication (diprosopus): CT, MR imaging, and MR angiography findings case report. Radiology 226: 210-213.