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Case Report

Diprosopus a Rare Craniofacial Malformation

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Abstract

Diprosopus is an extremely rare form of craniofacial malformation seen in newborns where there is duplication of face which may be partial or complete. The baby usually has a single trunk and normal limbs. We report a case of diprosopus admitted in our institute and analyze the anomalies and clinical problems and outcome of the case.

Keywords: *Diprosopus, facial duplication, neurulation*

Introduction

Diprosopus is a greek term meaning duplication of face. The patient typically has craniofacial duplication with normal trunk and limbs. It is a rare form of conjoined twins with a reported incidence of 1 case in 180,000 to 15 million births.[1] The most frequent type of conjoined twins is thoracopagus (32.7%), with joining at or near the sternal wall and contained viscera, and the rarest type is diprosopus (0.4). Two possible mechanisms leading to diprosopus has been proposed. First mechanism is possible cranial bifurcation of the notochord during neurulation. The bifurcation causes two vertebral axes and neural plates to develop alongside each other. Another proposal is an increase in the expression of the protein sonic hedgehog, which is essential for craniofacial patterning during development.[2] Advanced maternal age, polyhydramnios, and consanguineous marriage are considered high-risk factors for diprosopus. Prenatal diagnosis using ultrasonography, computed tomography (CT) scan, and magnetic resonance imaging (MRI) are possible. If diagnosis is made early in pregnancy, termination of pregnancy is sometimes considered as an option. Usually, diprosopus patients are stillborn, however, if not the prognosis is poor.

Case Report

A 20 days old, 36 weeks of gestation male baby born by normal vaginal delivery of a 36-year-old female presented with facial duplication and was admitted at Neonatal Intensive Care Unit of Gauhati Medical College. The patient had severe respiratory tract infection and malnutrition. Weight of the baby at birth was 3.4 kg and weight at the time of admission was 2.4 kg. Mother had another girl child of 3 year old and she was healthy. Head circumference was 36 cm, the baby had widely placed eyes, duplicated nose with three nasal openings, two oral cavity, two palate, two tongue, two maxilla, two mandible, duplicated pharynx, duplicated esophagus, and duplicated trachea. Plain CT scan Brain was done as shown in Figures 1 and 2 which revealed a cleft arising anterior to 3rd ventricle on both sides traversing anteriorly causing duplication of frontal lobes. Bilateral lateral ventricles were widely separated and thinned out; however, temporal and occipital horn appeared normal. Third ventricle was dilated, corpus callosum was not visualized suggesting complete absence, Sylvian fissures, and basal ganglia were lateralized but appeared normal. Posterior fossa contents including cerebellar hemisphere, vermis, and 4th ventricle were normal. The baby shown in Figures 3-5 succumbed to respiratory tract infection 48 h after admission.

Discussion

Diprosopus is a rare clinical entity with very few reported cases in literature and no reported cases from northeastern part of India. There is only about 35 reported cases in the literature.[3] It has a predominance of females over males 2:1. A complete duplication is associated with a high incidence of anomalies in the central

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nervous system, cardiovascular system, gastrointestinal system, and the respiratory systems, as well as cleft lip and palate.[4] The most accepted theory presently is that conjoined twins result from an embryological disturbance in the separation of the twins during the 2nd week of pregnancy (12–13 days), as a result of the abnormal splitting of the postimplantation blastocyte.[5] Recently, it has been postulated that conjoined twins result from the development of two independent notochords which were initially destined to become separate twins, but which were too close to develop independently.[6] In 1982, Barr classified duplication into three main forms (Barr, 1982):

I. Duplication of the eyes and nose with or without maxillary duplication by itself or with mandible duplication
II. Duplication of the nose with or without maxillary duplication
III. Duplication of the maxilla with or without mandible or pituitary duplication. He further described pituitary duplication in isolation but was uncertain regarding the existence of isolated mandibular duplication.

Later, Gorlin *et al*. (1990) created a classification scheme with an emphasis on oral duplication:

I. A single mouth with duplication of the maxillary arch
II. A supernumerary mouth laterally placed with a rudimentary mandible
III. A single mouth with replication of mandibular segments
IV. Diprosopus with or without anencephaly.

Risk factor for diprosopus includes familial tendencies, advance maternal age, previous history and polyhydramnios. Prenatal ultrasonography, CT scan, and MRI can detect diprosopus in cases with a high degree of suspicion. Most of the patients are stillbirth, and overall survival in live birth is poor due to multiple associated anomalies.

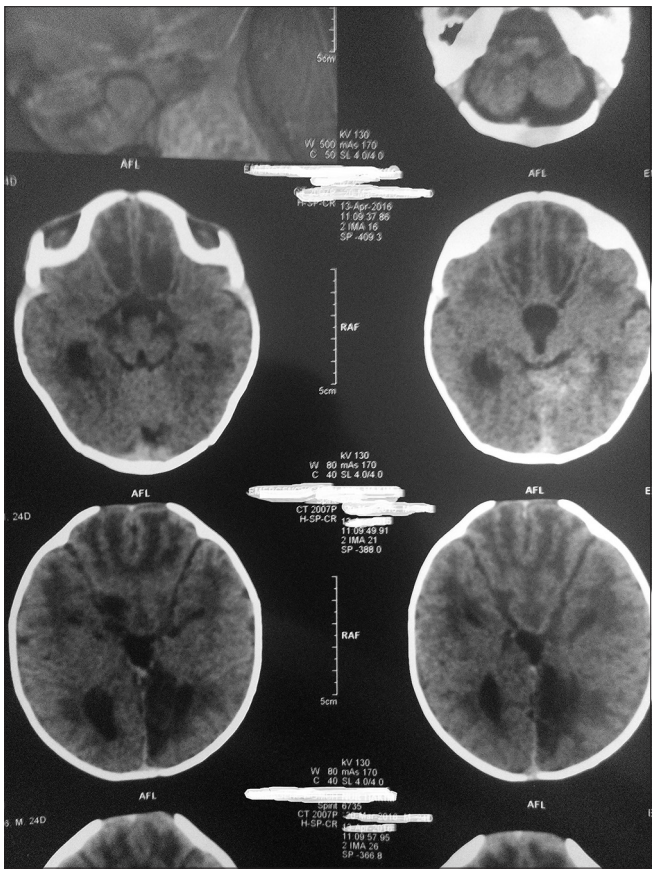

Figure 1: Computed tomography scan of the diprosopus case

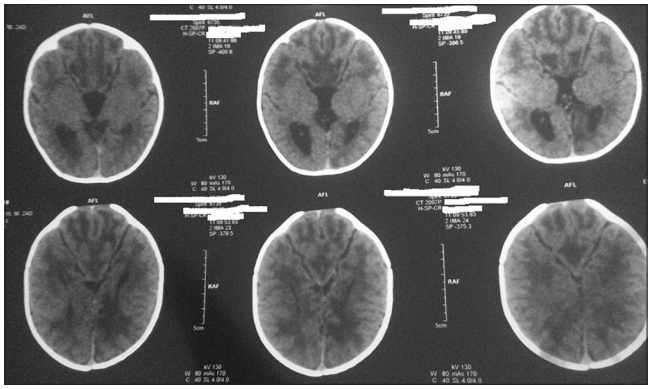

Figure 2: Computed tomography scan of the diprosopus case

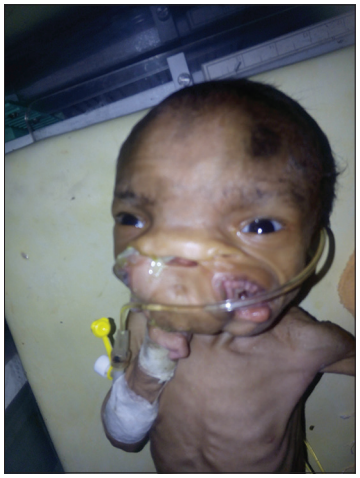

Figure 3: Case of diprosopus

Figure 4: Case of diprosopus

Figure 5: Case of diprosopus

Treatment options include surgical repair in selected incomplete variety and management of complications arising out of other anomalies.

Consent Ethical aspect

Prior consent from the patients authorized guardian was taken in writing after detailed explanation. Institutional approval was taken for the study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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